ORTHOPAEDICS

Principles and Their Application

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TO

MY WIFE

SARAH TURCK

Preface

Orthopaedic surgery has progressed from an immature limited specialty to a practice of scientific exactness encompassing a vast field of interrelated medical sciences. This book was born of a desire to compile scientifically accurate information relating to orthopaedic surgery and to formulate a method by which these facts are readily accessible.

Science implies truth. It is not opinion. A man who makes statements based upon opinion and tries to convince others that he is justified because he is entitled to his own opinion lacks the scientific approach. Scientific fact must be based upon documented irrefutable evidence. Only by insisting on truth and accuracy in investigative research can we progress in our diagnostic and surgical technics.

About 25 years ago, at the outset of my interneship training, the practice of orthopaedic surgery consisted of attempts to treat and rehabilitate patients affected by a variety of ill-understood conditions, including chronic osteomyelitis, poliomyelitic paralysis, "sciatica" and the like. Fractures, hand injuries and peripheral vascular disease came under the care of the general surgeon. The orthopaedic service seemed to be the least attractive to the interne. The literature was replete with opinionated, unsubstantiated reports and treatments which were admittedly empirical.

Nevertheless, a number of dedicated workers, each concentrating upon his own individual interest, labored to bring order out of chaos. In no small measure, tremendous impetus was derived from the efforts of the American Academy of Orthopaedic Surgeons. In certifying graduate students, the American Board of Orthopaedic Surgery recognized the importance of basic science studies. World War II afforded unlimited material for study and improvement of surgical technics. There remained only the task of collecting and organizing the facts so that the specialty could proceed on a firm footing.

The problem of sifting and correlating the information became apparent to me about 18 years ago. To increase my own efficiency and have ready access to accurate and up-to-date material, I prepared and maintained a classified file of information. First it was necessary to provide a foundation of basic scientific knowledge. Thus was evolved the sections on bone development, histology, physiology and basic pathology. Next there followed the recording of general conditions. Any disease peculiar to a certain location was classified within that particular region. The advantage of this procedure in a differential diagnosis is easily apparent.

Every effort was made to ascertain and record gross and microscopic pathology. The understanding of a disease process is a basic prerequisite to intelligent interpretation of the clinical picture, establishing the prognosis and formulating the treatment. It is a natural steppingstone in the investigation of etiology. The importance of histologic appearance and staining qualities of tissue is reflected in the enormous expenditure of time and effort in reproducing the colored photomicrographs in this book. It has always amazed me that pathologists everywhere confess their lack of knowledge about the musculoskeletal system. Descriptions in textbooks are inadequate. The orthopaedic surgeon was compelled to become a student of orthopaedic pathology and necessarily be self-reliant in making prognoses and surgical decisions. The histologic sections from which the photomicrographs have been prepared were obtained chiefly from the slides used for study by graduate students in the Orthopaedic Laboratory at Northwestern University Medical School.

This work constitutes an effort to record, between two covers, accurate scientific facts necessary to the study and the practice of orthopaedic surgery. The scope of the specialty extends far beyond that of 25 years ago. The study of gross anatomy, histology, pathology and physiology is fundamental to the armamentarium of the orthopaedic surgeon. He must comprehend architectural and engineer-

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PART ONE

The Basic Sciences

1

Development of the Skeleton

Within the first few weeks of life, the embryo passes through blastulous and the gastrulous stages and gradually begins to take shape, developing the head, the trunk and the external protrusions designated as limb buds. Between the ectoderm and the entoderm lies a diffuse, loose, cellular tissue, the mesenchyme, which differentiates into various connective tissue structures including bone, cartilage, fascia, muscles, etc. The earliest musculoskeletal structures are recognized as dense concentrations of mesenchymal cells which tend to take the shape of the bones of which they are forerunners. Each compact mesenchymal model then is converted into bone, either directly, e.g., cranial and facial bones, or indirectly by first being converted into cartilage which must be replaced by bone, e.g., the long bones.

Each musculoskeletal unit develops most actively during a specific period of early embryonic life. It is at this time that the development is susceptible to external toxic influences. Thus a specific congenital anomaly may be related to, for example, an attack of German measles occurring at a time during which that part is being energetically transformed.

CARTILAGE

As early as the 5th embryonic week, mesenchymal cells enlarge, become more compact and differentiate into a sheet of cells known as *precartilage*. Then matrix is laid down between the cells. The matrix contains fibrils which are peculiar to the type and the function of the cartilage. In *hyaline cartilage* the fibrils are not demonstrable by ordinary staining methods, so that the matrix appears clear and homogeneous. In *elastic cartilage* yellow elastic fibers are seen. In *fibrocartilage* heavier white fibers are deposited within the matrix. Cartilage increases in thickness by growth, both internally and externally. Internal growth is by multiplication of cartilage cells and production of new matrix. Peripheral growth takes place from the investing sheath, the *perichondrium*, whose inner cells are transformed into cartilage cells.

BONE

After the 7th embryonic week, bone first appears. This bone is of two types: membranous and cartilaginous. *Membrane bones* are those which form directly in membranous sheets, e.g., facial and cranial bones. *Cartilage bones* are those in which cartilaginous structures are first formed and then replaced by bone. Although the histogenesis of bone is identical in each instance, in the latter type the cartilage must first be removed before bone can be laid down.

Membranous Bone

The mesenchymal or connective tissue membrane first forms the original model of the facial and the cranial bones. At one or more central points of the membrane *intramembranous ossification* begins. These ossification centers are characterized by the appearance of osteoblasts which lay down a meshwork of bony trabeculae spread radially in all directions. The mesenchyme at the periphery differentiates into a fibrous sheath, the periosteum, the undersurface of which differentiates into osteoblasts, which in turn deposit parallel plates of compact bone, the lamellae. This is *periosteal ossification*, by which the inner and the outer tables of the skull are formed. Trabeculae are mainly arranged along lines of greatest stress.

Cartilage Bone

A cartilaginous model of the structure precedes destruction of cartilage and its replacement by bone. Two processes are involved:

lage disintegrates and is destroyed by invasion of vascular tissue from the perichondrium. At the same time the invasive budlike mass gives rise to osteoblasts which deposit new bone at many points and even upon the calcified cartilage. This spongy bone formation continues to replace the cartilage, extending proximally and distally.

Periosteal Ossification. At the same time as the process of spongy central bone formation is going on, the inner layer of perichondrium, now more appropriately named the periosteum, is laying down parallel layers of compact bone.

The process of endochondral ossification continues throughout the growth period by persistence of a layer of cartilage near the epiphyses and is responsible for growth in length of the structure. Periosteal ossification contributes to growth in thickness of the structure. These processes are described in detail under Histogenesis.

JOINTS

Joints occur where bones meet and are of two types: *synarthroses,* in which little movement is allowed, and *diarthroses,* in which movement is free.

The synarthrosis is formed by differentiation of mesenchyme into a uniting layer of connective tissue, the *suture or syndesmosis*; cartilage, the *synchondrosis*; or bone, the *synostosis.*

The diarthrosis is characterized by a joint cavity which arises by a cleft in the mesenchyme. The capsule forms from the dense external tissue which is continuous with the periosteum. The cells on the inner surface of the capsule flatten into a false epithelium called the *synovial membrane.* Ligaments or tendons which apparently course through the cavity represent secondary invasions covered with synovial membrane reflected upon them and therefore are really external to the cavity. An articular disk is a fibrocartilaginous plate formed from mesenchyme midway in the cavity.

MORPHOGENESIS OF THE AXIAL SKELETON

The notochord is the primitive axial support. Mesenchymal tissue, designated as sclerotomes, migrate toward the notochord and come to lie in paired segmental masses alongside the notochord. Each sclerotomic mesenchymal mass is separated from similar masses before and behind by the intersegmental arteries. Each sclerotome then differentiates into a caudal compact portion and a cranial less-dense half. The denser caudal half then unites with the looser cranial half of the next succeeding sclerotome to form the substance of the vertebra. Both the condensed and the looser portions grow about the notochord to form the *body* of the vertebra. From the denser (now cranial) half dorsal extensions pass around the neural tube to form the *vertebral arch,* and paired ventrolateral outgrowths form the *costal processes* or forerunners of the ribs. The mesenchymal tissue in the intervertebral fissure gives rise to the *intervertebral disk.* The nucleus pulposus in the disk constitutes the remnant of the notochord. The two parts of sclerotomes in joining enclose the intersegmental artery which therefore passes through the center of the vertebral body. In the 7th week, centers of chondrification appear, 2 in the vertebral body and 1 in each half of the vertebral arch. These 4 centers enlarge and fuse into a complete cartilaginous vertebra. Vertebral ossification starts in the 10th week. A single center in the body and 1 in each half of the arch appears, but union is not completed until several years after birth.

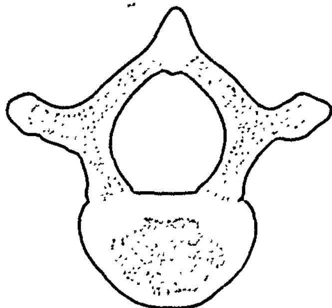


FIG. 4. Ossification centers in vertebra.

Continued growth in length of the body

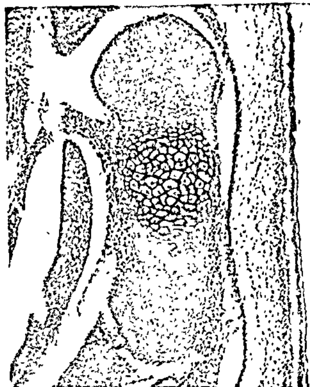


FIG. 1 Early fetal anlage of a long bone. It is composed of mesenchyme at the center of which the cells become rounded and assume the appearance of chondrocytes. Later, the peripheral mesenchyme will give rise to vascular tissue which will invade the calcified cartilage and replace the latter with bone.



FIG. 2. Ossification of a fetal cartilaginous long bone. The cartilage cells at the center of the calcified cartilage have become enlarged, and the matrix is sparse. A bone collar has formed about this level and is gradually replacing the cartilage.

(1) ossification centrally within the cartilage, or *endochondral ossification*, (2) and peripherally beneath the perichondrium (or periosteum), or *perichondrial or periosteal ossification*.

Endochondral Ossification. In the center of the cartilaginous precursor, the cells enlarge *and become arranged radially*. Lime salts are deposited in the matrix. This calcified carti-

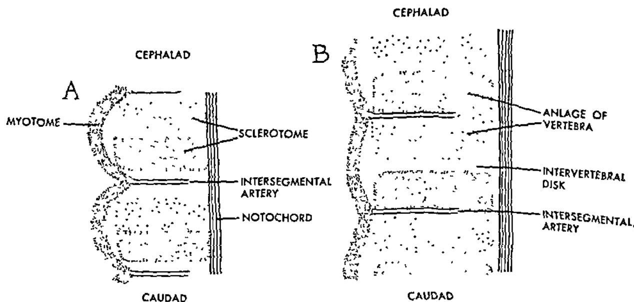


FIG. 3. Early stages of differentiation of vertebrae (Redrawn from Arey, L. B.: Developmental Anatomy, Philadelphia, Saunders)

lage disintegrates and is destroyed by invasion of vascular tissue from the perichondrium. At the same time the invasive bud-like mass gives rise to osteoblasts which deposit new bone at many points and even upon the calcified cartilage. This spongy bone formation continues to replace the cartilage, extending proximally and distally.

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Continued growth in length of the body

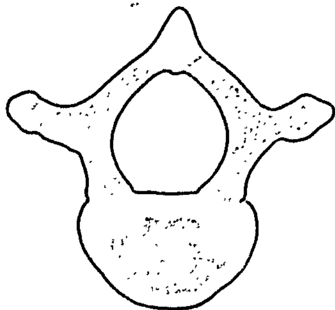


Fig. 4. Ossification centers in vertebra.

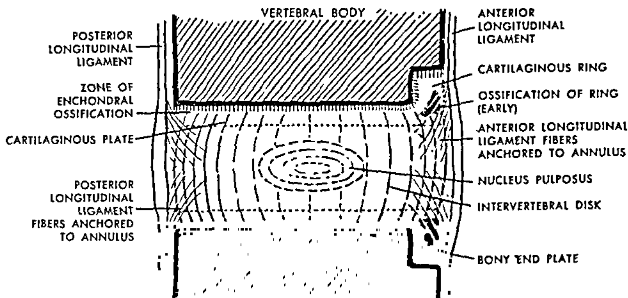


FIG 5 Sagittal section through adjacent vertebral bodies during ossification of the cartilaginous ring (Redrawn from Schmid, P.: Zur Entstehung der Adoleszentenkyphose, Deutsche Med Woch 25:798)

takes place by endochondral ossification at the cephalad and the caudad epiphyseal plates. About the rim of the superior and the inferior surfaces a prominent ring of cartilage exists to which is attached the fibers of the longitudinal ligament of the spine. This does not participate in growth. Gradually, it develops secondary ossification centers which are triangular in cross section but actually skirt the rim of the body. Eventually, this center appears as a line parallel with the upper and the lower surfaces of the body and resembles a plate. The term "plate" is reserved for the growth cartilage which intervenes between the ring and the main body of bone. The secondary centers fuse with the main body by the age of 17. The central artery can be seen up to 6 years of age, after which it is obliterated. It may persist beyond this time in certain conditions, as in Scheuermann's disease. An exception in development of the vertebra occurs in the atlas. The body differentiates typically but soon is taken over by the epistropheus (axis) serving as a peglike extension (dens) of the latter, about which the atlas rotates. The atlas is left as a ring. The sacral and the coccygeal vertebrae represent types with reduced vertebral arches. The sacral vertebrae eventually fuse into a single mass. The coccygeal vertebrae exist as rudimentary structures. The entire spine at birth displays one continuous curve convex posteriorly. As the posture of the erect position is assumed after the first year, secondary forward curves develop at the cervical and the lumbar regions. Finally, the lordosis in the cervical and the dorsal regions is balanced by the kyphosis in the thoracic and the sacral regions.

The original union of the costal process with the vertebra is replaced by a joint for the head of the rib. The center of ossification appears at the angle of the rib However, the distal ends of the long ribs always remain cartilaginous. In the neck the ribs are represented by their tubercles, which are fused with the transverse processes and their heads with the bodies; between these processes is an interval, the *transverse foramen*, through which the vertebral arteries course. When the costal processes are overdeveloped in the cervical region, a supernumerary rib is formed and may lead to compression of nerve structures.

The sternum originates from the junction of 2 bars of ventrolaterally placed mesenchyme, which initially have no connection with the ribs or with each other.

MORPHOGENESIS OF THE APPENDICULAR SKELETON

This consists of a cranial and a caudal internal support or girdle and the skeleton of

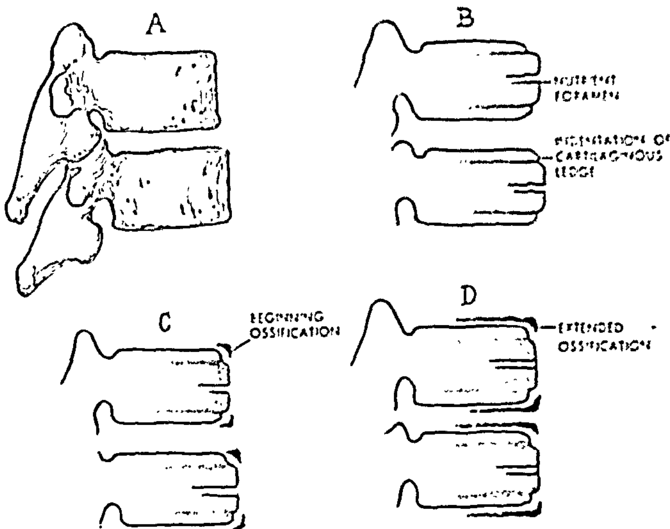


FIG. 6. The appearance of normal thoracic vertebrae in children at various ages. (A) Normal thoracic vertebrae, fully developed, lateral view. (B) Lateral x-ray view of vertebral bodies in children under 6 years of age (diagrammatic). (C) Lateral view in children, 6 to 9 years of age. Ossification starting in the cartilaginous ring most visible anteriorly. (D) At 9 to 15 years of age. Ossification more extensive in the ring and progresses posteriorly. The nutrient foramen normally is obliterated after age 6.

free appendages attached to them. The appendicular skeleton is derived directly from the unsegmented somatic mesenchyme. Definite masses are formed at the sites of the future pectoral and pelvic girdles and limb buds. This is followed by the sequence of bone development through cartilaginous and osseous stages.

The *clavicle* is the first bone of the skeleton [illegible] appear.

The *scapula* is a single plate with two chief centers of ossification and several epiphyseal centers which appear later. An early primary center forms the body and the spine. The other, after birth, gives rise to the coracoid process.

The *humerus*, the *radius* and the *ulna* all ossify from a single primary center in the diaphysis and an epiphyseal center at each end. Additional epiphyseal centers are constant at the lower end of the humerus. Each carpal bone ossifies from a single center. The metacarpals ossify from a single primary center and an epiphyseal center.

At first, the *cartilaginous plate of the pelvis* lies perpendicular to the vertebral column. Later, it rotates to a position parallel with the vertebral column and in relation to the first 3 sacral vertebrae. Three main centers of



FIG. 7. (A) Origin of a limb bud. Development of the upper limb bud and histogenesis of the humerus are shown. The limb bud originates as a small elevation of the body wall and at first consists of a condensed mass of proliferating mesoblastic cells. Within a few days, a central condensation of mesoblasts takes place. This is the *skeletomuscle condensation*, so-called because separate muscle and skeleton cannot be identified. At the same

(*Continued on facing page*)

ossification appear for the ilium, the ischium and the pubis. The 3 elements join at a cup-shaped depression, the acetabulum, the articulation for the head of the femur.

The development of the femur, the tibia, the fibula, the tarsus, the metatarsals and the phalanges corresponds to the bones of the upper extremity.

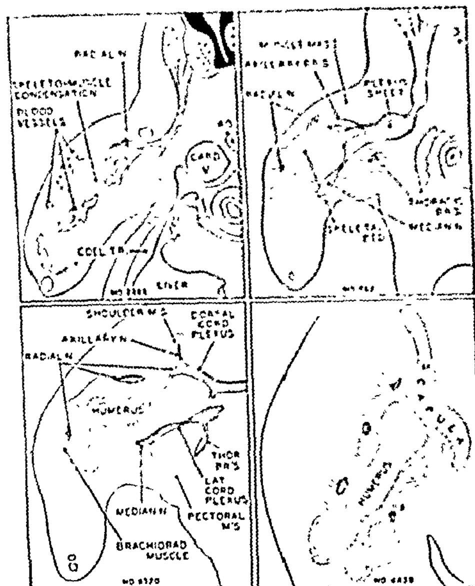


FIG 7 (*Continued from facing page*)
time, a broad sheet of branching nerve trunks from the cord and the spinal ganglia stream into the base of the arm bud (*top, left*). A few days later, muscular and skeletal condensations become distinct, and massive nerve trunks enter the center of the muscle condensations (*top, right*). In the next section (*bottom, left*), definite muscle groups can be identified containing conspicuous nerve trunks, and major branches of the brachial plexus are obvious. At the same time, the central part of the skeletal condensation is being transformed into cartilage (this tissue appears lighter in color). The cartilage of different bones is deposited separately so that in the last section (*bottom, right*) these central cartilaginous cores within the skeletal bed acquire the shape of the bones of which they are forerunners. These sections represent developmental periods of about 2 days each.
(B) These outline drawings correspond to these sections in (A).
(Streeter, G. L.: Developmental horizons in human embryos [Pub. 583], Contrib. Embryol. 33:149. From the Carnegie Institution of Washington.)

2

Histology of Bone

Bone is a specialized connective tissue with a calcified collagenous intercellular substance for skeletal support of the body. It is either spongy (cancellous) or compact in structure. Spongy bone consists of intercrossing and connecting bars of varying shapes and thickness, between which are spaces filled with bone marrow. Compact bone is a continuous hard mass whose spaces are microscopic in size. Both types exist in almost every bone.

Typical Long Bone (e.g., femur). The *diaphysis* (shaft) consists of a wall of compact bone enclosing a large cylindrical bone-marrow cavity.

The *epiphysis* (end of bone) consists of spongy bone with a thin outer wall of compact bone. In a growing animal, the *epiphyseal cartilage plate*, from which longitudinal growth occurs, lies between the epiphysis and the diaphysis.

The *metaphysis* is the spongy bone directly beneath the epiphyseal plate and is the most recently formed bone arising out of the growth process at the plate.

Flat Bones of the Skull. These are composed of inner and outer layers (tables) enclosing spongy bone (diploe).

Small Bones (e.g., carpals) are of simple construction, an outer wall of compact bone enclosing spongy bone.

Periosteum is a modified connective tissue which covers the bone.

Endosteum, a similar tissue, lines the marrow spaces.

GENERAL MICROSCOPIC APPEARANCE

Layers (lamellae) of calcified *bone matrix or interstitial substance* are seen as clear material, although by appropriate staining methods they are fibrillar in nature. Cavities (*lacunae*) within the interstitial substance are occupied by bone cells (*osteocytes*). All lacunae are connected by a tremendous number of minute canals (*canalicules*) which penetrate the hard substance, branch abundantly and anastomose. Osteoblasts (bone-forming cells) and osteoclasts (bone-absorbing cells) line the surfaces of bone.

Osteoblast. Osteoblasts line the surface of actively growing bone in great numbers. The body is from 15 to 20 microns in diameter, contains a large nucleus and one fairly large nucleolus. The cytoplasm stains intensely with basic aniline dyes. Phosphatase is contained within it. Osteoblasts are often connected with each other by thin cytoplasmic processes.

Osteocyte. This is an osteoblast which has become imprisoned within the bone matrix. It has a faintly basophilic cytoplasm, large oval nucleus with large chromatin granules and one or more nucleoli. The cytoplasms of both osteocytes and osteoblasts contain spherical granules stainable with periodate-leukofuchsin, suggesting a common origin.

Lacunae. These cavities are flat and oval. Projections from the cell bodies enter fine apertures in the walls of the lacunae.

Osteoclast. This consists of a multinucleated giant cell varying in size and number of nuclei. The cytoplasm is pale-staining, acidophilic and foamy. The nuclei are poor in chromatin, but each has a [illegible] osteoblasts or from stromal cells of the marrow.

Interstitial Substance. By ordinary staining methods, the interstitial substance appears to be homogeneous. However, *osteocollagenous fibers* (ossein) similar to collagenous fibers of loose connective tissue are contained within the substance and are demonstrable by silver impregnation. They are collected in bundles and are united by an amorphous binding substance. This amorphous substance is barely stainable but becomes more vividly stained as pink *osteoid* when bone is being deposited rapidly. It is in this organic substance that

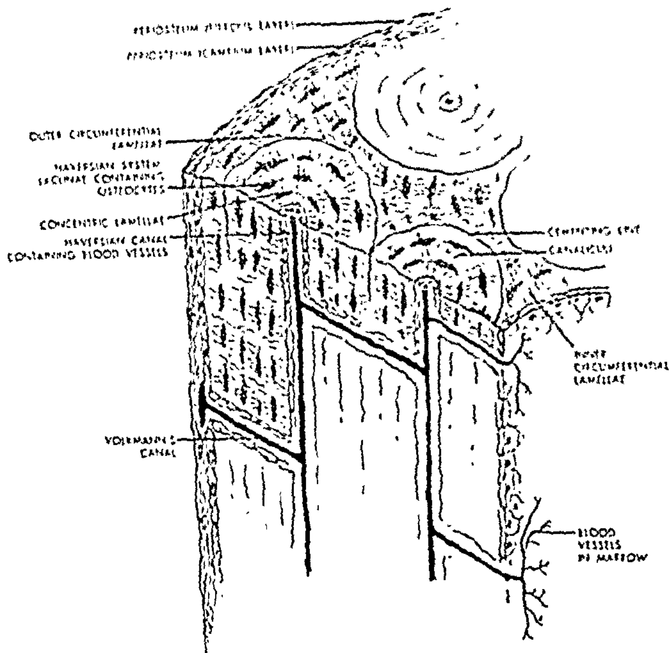


Fig. 8. Diagrammatic representation of a cross section and a longitudinal section of the cortex of a long bone. Note the haversian systems running longitudinally. Volkmann's canals constitute connecting channels between periosteal and haversian and bone marrow blood vessels. (Redrawn from Ham, A. W.: Histology, Philadelphia, Lippincott)

the minerals are deposited. The hard interstitial substance is composed of water, the organic framework and inorganic salts. The mineral salts consist mostly of submicroscopic crystals of hydroxyapatite $[Ca_3(PO_4)_2]3$ $Ca(OH)_2$ or the closely related *hydrated* tricalcium phosphate. The apatite lattice is determined by x-ray spectograms. Sometimes radium and fluoride are substituted in the lattice. Other cations, especially magnesium and sodium, and anions, as carbonate and citrate, are found. The substances are believed to be on the surfaces of the crystal lattice. The bone salt composes 65 per cent of the adult dry bone. This is reduced to 30 to 35 per cent in rickets and osteomalacia.

The organic constituent of bone is the *bone collagen or ossein.* When boiled, gelatin results.

A weak acid, such as glycerine, will remove the inorganic salts and leave the original structure grossly and microscopically.

STRUCTURE OF BONE

The *cortex* of a bone is composed of compact bone, while the *medulla* contains cancellous, or spongy, bone. Spongy *bone* is made up of a loose network of trabeculae of bone which are interconnected but generally ar-

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Fig. 9. Cortex of long bone, showing appearance of haversian canals in longitudinal section (A) and in cross section (B).

ranged along lines of maximum stress or tens. The *trabeculae* are made up of a varying number of adjoining bone plates. Osteocytes within the lacunae communicate with each other by *canalicules.*

Compact bone consists of *lamellae* whi are regularly arranged about branching a anastomosing canals through which nutrie vessels pass. These *haversian canals* commu cate with the outer surface of the bone or t medullary cavity through *canals of Volkman*

The typical *haversian system* is the ba *structure of compact bone. It consists* lamellae concentrically arranged about t haversian canal. Most haversian systems a directed in the long axis of the bone. Ther fore, in cross section the canals appear small rounded openings, and the lamellae circles; in longitudinal section the canals a pear as long slits Large numbers of canali ules pass radially from the canal to the lacun and intercommunicate with each other. The function is supposedly for diffusion of nutrie fluids. Compact bone is made up of large nu bers of haversian systems between which a *interstitial* or *ground* lamellae. These latt are the remains of haversian systems whi are only partly destroyed. On the outer a internal aspects of the compact bone a lamellae which are arranged circumferential in relation to the main bone. These are t *basic or circumferential lamellae.* These a

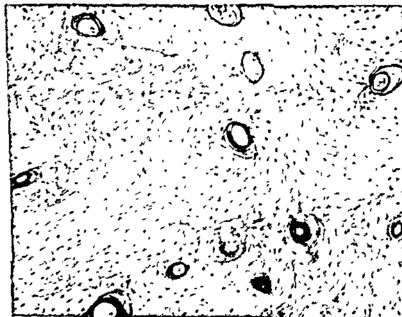


Fig. 10. Cross section of cortex of a long bone. The haversian systems are well displayed.

penetrated by the *canals of Volkmann* through which nutrient vessels enter the bone to reach the vessels in the haversian canals. They contain large vessels and are not surrounded by concentrically arranged plates. *Sharpey's fibers* are thick bundles of collagenous fibers which pass from the periosteum into the basic external circumferential lamellae. They fix the periosteum firmly to the surface of the bone, particularly where tendons and muscles attach, where large blood vessels and nerves enter the bone, and at the epiphyses of long bones.

The *periosteum* consists of dense connective tissue containing blood vessels. Its deepest stratum, the *cambium layer*, is more loosely arranged and contains spindle-shaped cells and a network of thin elastic fibers. In the adult the periosteum is not osteogenic until necessity arises, as after a fracture, whereupon osteoblasts appear in the cambium layer.

The *endosteum* is a thin layer of connective tissue which lines the walls of bone cavities, including marrow spaces. It can become osteogenic and hematopoietic.

HISTOGENESIS OF BONE

In embryonic life most of the skeleton is composed of cartilage which is absorbed and replaced by bone. This is called *endochondral ossification*. When bone is formed directly from connective tissue without the intervening stages of cartilage formation and reabsorption, it is called *intramembranous ossification*.

Intramembranous Ossification

The original connective tissue is a rather loose tissue with delicate fibrils between which is an amorphous fluid substance. The elliptical connective tissue cells differentiate into osteoblasts by increase in size and become polyhedral with numerous processes. Bars of dense [illegible] ubstance appear, [illegible] amorphous fluid [illegible] steoblasts, which are now called [illegible] which line the surface of the developing bone in large numbers. New osteoblasts are constantly formed by transformation of other connective tissue cells. A network of [illegible]

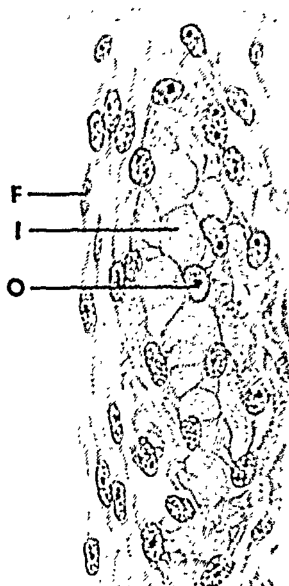


FIG. 11. Intramembranous bone formation. (F) Fibroblasts. (I) Homogeneous interstitial bone substance, collagenous fibrils no longer visible. (O) Connective tissue cells which have developed processes to become osteoblasts and later osteocytes.

branching interconnecting [illegible] formed, between which [illegible] sue which is very [illegible] multiplying cells. [illegible] within spaces [illegible] tissue becomes [illegible] lying osteoblasts [illegible] of [illegible] [illegible]



PLATE 1. Endoch dral ossification. capillary loops are s entering and destr ing the piled-up ca lage cells. At the l is the layer of pro erating chondrocyt Proceeding toward right, one observes c cified cartilage whi becomes envelop with osteoid laid dov by a profusion of oste blasts. The earliest a pearance of matu bone is at the extren right of the section.

tion of the cartilage, invasion by capillaries, resorption of the calcified cartilage, and at the same time formation of new bone by the same method as in intramembranous ossification. Endochondral ossification takes place continuously at the epiphyseal plates of long bones until growth in length of these bones is complete This is the most advantageous site for study of this process.

Irregularly distributed groups of cartilage cells multiply and become arranged in orderly columns of flattened cells separated by thin capsules. The columns are separated by wide bands of interstitial substance. In the next zone distally, the cells swell and become vacuolized and degenerate. These are the *vesicular or hypertrophic cartilage cells*. The adjacent matrix calcifies and stains deeply, forming the *zone of provisional calcification*. Loops of blood vessels with accompanying connective tissues penetrate the cartilage by invading and destroying the cells. The vascular channel is formed adjacent to the calcified matrix. The elongated connective tissue cells become

is reabsorbed at the same time. Eventually, the bone thickens as it proceeds distally and contains small remnants of calcified cartilage as evidence of its endochondral origin.

If a deficiency of bone minerals exists, th cartilage matrix fails to calcify and cannot b removed. Multiplication of cartilage cells con tinues, and the epiphyseal plate becomes ver thick. However, pink-staining uncalcified osteoid tissue is laid down on the cartilaginous matrix in large amounts. This is the picture in rickets or osteomalacia.

REMODELING OF BONE

Throughout life bone is constantly being resorbed and reformed, thereby being essential to form and size of the bone and forming a source of supply of calcium to the body. Osteoclasts are almost always seen in areas of bone destruction and disappear when bone formation is the primary activity. The osteoclast can be found in a deep indentation (Howship's lacuna) in a trabeculum suggesting erosion, but actual phagocytosis by the cell cannot be demonstrated. The rate of bone reconstruction is to some degree related to the available ionized plasma calcium. When in the adult the new osseous tissue is formed but remains uncalcified as osteoid tissue, oste malacia results.

Some regions of spongy bone are cor verted into compact bone. In an area c marrow, osteoblasts lay down a periphera circumferential bone plate. Next, anothe plate is laid down within the first plate, the successive concentric layers are add

marrow space with its blood vessels becoming progressively narrowed until a *primitive haversian system* is formed. Within the area, destruction of bone starts, associated with the presence of osteoclasts, and cavities filled with embryonic bone marrow and blood vessels result. Then again osteogenesis starts, and another haversian system is formed. This

after E.B Patterson.

FIG. 12. Development of a typical long bone, ... model. (b) Periosteal bone collar appears. (c) ... (d) Further development of calcified cartilag... enters, resorbs calcified cartilage, and new bone is laid down toward either extremity of the model (f) Endochondral ossification further advanced, bone increased in length. (g) Blood vessels and mesenchyme enter upper epiphyseal cartilage (h) Development of epiphyseal ossification center. (i) Ossification center develops in lower epiphysis. (j and k) The lower and then the upper epiphyseal cartilage plates disappear, bone ceases to grow in length, a continuous bone marrow cavity traverses the entire length of the bone, and blood vessels of diaphysis, metaphysis and epiphysis intercommunicate. (Redrawn from Maximow and Bloom: Textbook of Histology, Philadelphia, Saunders)



PLATE 1. Endochondral ossification. The capillary loops are seen entering and destroying the piled-up cartilage cells. At the left is the layer of proliferating chondrocytes Proceeding toward the right, one observes calcified cartilage which becomes enveloped with osteoid laid down by a profusion of osteoblasts. The earliest appearance of mature bone is at the extreme right of the section.

tion of the cartilage, invasion by capillaries, resorption of the calcified cartilage, and at the same time formation of new bone by the same method as in intramembranous ossification. Endochondral ossification takes place continuously at the epiphyseal plates of long bones until growth in length of these bones is complete. This is the most advantageous site for study of this process.

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Some regions of spongy bone are converted into compact bone. In an area of marrow, osteoblasts lay down a peripheral circumferential bone plate. Next, another plate is laid down within the first plate, then successive concentric layers are added, the

3

Histology of Cartilage

Cartilage is a specialized connective tissue which forms the temporary skeleton in the embryo as a model upon which bones develop. It persists in the adult animals in joints, respiratory passages, ribs and ears. The quantity of intercellular substance is large in contrast with the cavities within it containing the cells, the chondrocytes.

HYALINE CARTILAGE

This is the most widespread type, covering the articular surfaces of bones, forming the anterior rib portions and existing within the respiratory passages. It is flexible, elastic, bluish-white and opalescent. The cells are mainly spherical, although near the surface they are flattened in a plane parallel with the surface. In deeper layers they may be hemispherical or angular. They completely fill the cavity. The cytoplasm contains long mitochondria, vacuoles, fat droplets and glycogen. The vacuoles may be large and distend the cell. The nucleus contains several spherical nucleoli. Mitotic figures are almost never found. The cells are aggregated into compact groups irregularly placed. A group is formed by rapid multiplication within a cavity. Or they may be assembled into columns of flattened cells where they proceed toward advancing bone formation at the epiphyseal plate. The interstitial substance is homogeneous except about the cell cavities, where it is different staining and occasionally exhibits concentric striations, appearing as *cartilage capsules.* Collagenous fibers within the interstitial substance are demonstrable by silver impregnation methods or by digesting the tissue with trypsin which does not affect the fibers.

Chondromucoid, a glycoprotein, is contained within the interstitial substance and is responsible for the basophilic staining. On hydrolysis it yields *chondroitin* sulfate.

Cartilage has no blood vessels except an occasional one passing through to other tissues. Nutritive tissue fluids from the perichondrium or the joint cavities permeate the interstitial substance.

ELASTIC CARTILAGE

This differs from hyaline cartilage in that the interstitial substance is penetrated in all

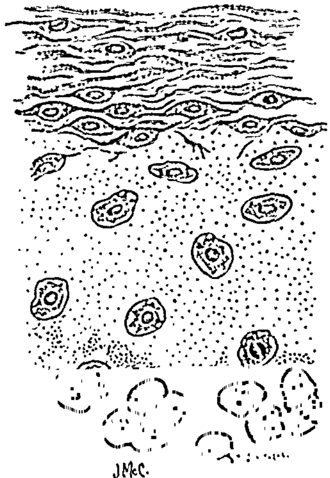


Fig. 13. Development of hyaline cartilage from perichondrium. The fibroblasts enlarge into chondroblasts and become encapsulated. Columns of isogenous cells appear in the deeper layers. The matrix stains more intensely immediately adjacent to these latter cells. (After Gordon)

process of destruction and reconstruction continues throughout life. Basic or circumferential lamella laid down by periosteum and endosteum likewise undergo destruction and replacement.

DEVELOPMENT OF THE WHOLE BONE

The long bone is preceded by a model of cartilage covered externally by a perichondrium of embryonic connective tissue. The perichondrial cells adjoining the cartilage become osteoblasts and lay down membranous bone in a ring encircling the diaphysis, the *periosteal bone band or collar.* This is a spongy network of bone. At the same time the cartilage tissue in the diaphysis changes. The cells swell and the interstitial matrix becomes calcified and thinner. Blood vessels invade from the periosteum through the interstices in the periosteal bone collar and enter spaces left by destruction of the cartilage cells. These spaces become filled with embryonic bone marrow. Osteoblasts appear and lay down new bone on a lattice of absorbing calcified cartilage. The cartilage model increases in size by continued growth of cartilage at the epiphyses.

Eventually, ossification centers appear in the epiphyses by the same process of endochondral ossification. Only the cartilage cells between epiphysis and diaphysis remain and continue to multiply for growth in length of shaft. When growth ceases, the cartilage plate or disk is replaced by bone (closure of the epiphyses). The first bone formed is spongy and found in large amounts in the metaphyseal area. Distally, as reconstruction proceeds, more compact bone forms

In short bones developing in cartilage, endochondral ossification starts centrally and progresses peripherally. The perichondrium then becomes periosteum and lays down periosteal bone, which becomes compact.

Flat bones which develop in cartilage (scapula) develop by periosteal ossification followed by endochondral ossification as in long bones.

Bones of the skull and most facial bones develop by intramembranous ossification.

The lower jaw has a special mode of ossification. It is formed in the embryo by cartilage (Meckel's cartilage) but does not ossify. Instead, the connective tissue covering lays down surface bone, and later the cartilage is absorbed.

BIOCHEMICAL CONSIDERATIONS

It is presently accepted that the mineral component of bone is a mixture of tricalcium phosphate and calcium carbonate rather than complex compounds of the apatite series.[1] The calcium content is dependent upon many factors, such as acid-base balance. For example, a slight decrease in pH (acidosis) will increase solubility of calcium carbonate and cause bone demineralization. These factors affecting calcium deposition and removal are described in the section on Physiology.

Only ionized mineral elements which compose only a portion of blood calcium and phosphorus can be utilized for ossification. The ionized calcium in the blood rises in acidosis and decreases with the rise in blood proteins. Precipitation of tricalcium phosphate in bone matrix takes place under the influence of glycogen, phosphorylase and phosphatase. Glycogen is split into phosphoric esters by phosphorylase. The esters in turn are hydrolyzed by phosphatase to provide inorganic phosphates. A concentration of phosphorus ions exceeds the product of solubility of calcium phosphate, and precipitation ensues. Next the calcium salts are fixed in the protein matrix. A proteinase secreted by white cells of the bone marrow splits the polypeptide chains of bone protein. Certain acid and base groups are liberated and combine with calcium and phosphorus ions to form calcium phosphoproteinate.

When calcifiable osseous tissue forms but is as yet uncalcified it is termed *osteoid,* a substance staining a light pink by hematoxylin and eosin stain. Osteoblasts contain phosphatase and are rich in ribose nucleic acid for formation of bone matrix.

[1] Cartier, P: Present state of biochemical problem of ossification, Rev orthop 33 272, 1947.

3

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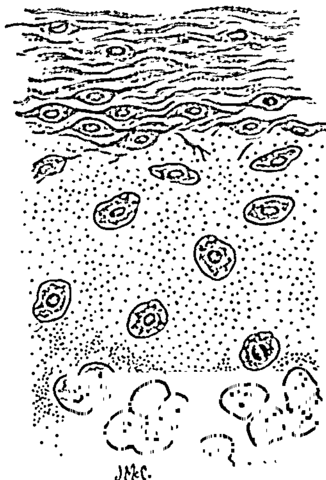


FIG. 13. Development of hyaline cartilage from perichondrium. The fibroblasts enlarge into chondroblasts and become encapsulated. Columns of isogenous cells appear in the deeper layers. The matrix stains more intensely immediately adjacent to these latter cells. (After Gordon)



FIG 14 Elastic cartilage (human ear).

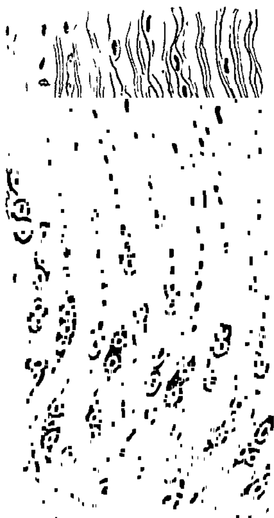


FIG. 15. Fibrocartilage. This is observed at osseotendinous junctions. Note the transformation of rows of tendon cells at the top into cartilage cells surrounded by deeply staining cartilaginous matrix. (Redrawn from Maximow and Bloom: Textbook of Histology, Philadelphia, Saunders)

directions by heavily staining branching elastic fibers which give the cartilage its yellow color, elasticity, flexibility and opacity. It occurs chiefly in the ear and other regions unrelated to the interest of the orthopaedic surgeon.

FIBROCARTILAGE

This differs from hyaline cartilage by the presence of thick, compact bundles of collagenous fibers within the interstitial substance. These bundles are arranged parallel with each other, separated by clefts in which the encapsulated cells are squeezed. Fibrocartilage appears to be a transitional tissue between hyaline cartilage and connective tissue and as such occurs in special situations as, for example, where the articular cartilage is connected to the dense connective tissue of capsules or ligaments of joints. It also occurs in the intervertebral disks, certain articular cartilages, the symphysis pubis, the ligamentum teres and in the attachment of certain tendons to bones.

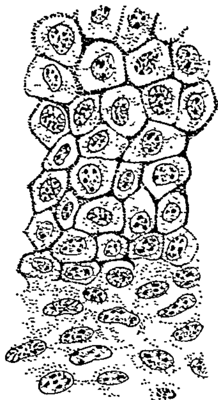


FIG. 16. Development of cartilage from mesenchyme. The cells of the mesenchyme (below) become rounded, encapsulated and separated by matrix (above).

HISTOGENESIS OF CARTILAGE

In the mesenchyme the cells become rounded, and the collagenous fibrils in the intercellular substance become enclosed by basophilic intercellular substance. The cells accumulate vacuoles, frequent mitoses occur, and daughter cells in a group are separated only by a thin partition. A thin, shining layer, the capsule, appears about the cell cavity and represents most recently formed intercellular substance. The mesenchyme about the cartilage model forms a connective tissue layer covering, the perichondrium. A constant transformation of these layers into cartilage occurs during embryonic life and through adulthood. The collagenous fibers are acidophilic flat bundles. These are surrounded by basophilic cartilaginous intercellular substance, while, at the same time, the fibroblasts lose their spindle shape and change to spherical cells, the chondrocytes, surrounded by capsules. This is *appositional growth* of cartilage as contrasted with interstitial growth wherein the cells within the cartilage multiply within their capsules and add to the surrounding matrix. Most of the embryonic cartilage of the skeleton is later replaced by bone.

REGENERATION OF CARTILAGE

When cartilage is injured, the cells become degenerate and necrotic. New connective tissue grows in from the neighboring area and fills the defect. The fibroblasts become rounded and encapsulated. New interstitial substance surrounds the fibrils, and the matrix becomes basophilic and homogeneous. Therefore, new cartilage forms by metaplasia of connective tissue. This occurs particularly under the influence of external forces, friction and pressure combined.

CARTILAGE CHANGES IN GROWTH

Before cartilage can be replaced by bone, the matrix must calcify and disintegrate, providing the necessary minerals for ossification. At the same time, the chondrocytes die. The chain of events is as follows. The cartilage cells proliferate, increase in size and advance toward the zone where ossification will occur. These multiplying cells become arranged in a large group (as in a vertebra or an os calcis) or in columns of cells separated by wide bands of matrix (as in the epiphyseal plate of a long bone). As the cells hypertrophy, they contain an increasingly larger amount of glycogen which will provide phosphorus for matrix calcification. In addition, the cells produce large quantities of alkaline phosphatase. Then, under the influence of an enzyme phosphorylase, the glycogen breaks down, forming hexose-phosphoric esters. These organic compounds are hydrolyzed by alkaline phosphatase to liberate inorganic phosphorus. The matrix then becomes calcified, and the chondrocytes lose their glycogen. The cells degenerate as vascular osteogenic granulation tissue penetrates the capsules, and ossification proceeds as the calcified matrix is resorbed.

PHYSIOLOGIC FACTS

When a rat is placed on a protein deficient diet, the width of the epiphyseal cartilage narrows. Deficiency of vitamin D, calcium, or phosphorus reduces calcification and ossification so that cartilage proliferation continues unchecked, resulting in marked widening of the epiphyseal growth plate. Diminution of anterior pituitary growth hormone reduces cartilage growth activity so that the cartilage plate is thinned. This is the picture in hypophysectomized animals. When growth hormone is injected into the animal, the cartilage resumes its normal growing appearance. Cartilage growth may also be reduced by thyroidectomy in animals; it is restored by injection of thyroxin. Cartilage is destroyed by irradiation with resultant retardation of growth.



FIG 14 Elastic cartilage (human ear).

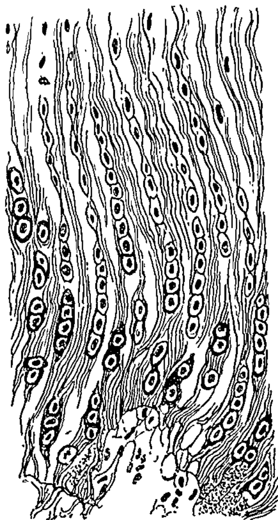


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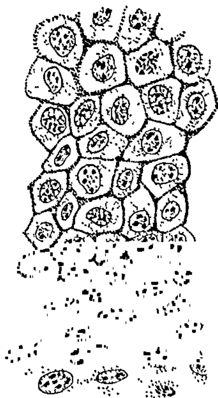


FIG. 16. Development of cartilage from mesenchyme The cells of the mesenchyme (below) become rounded, encapsulated and separated by matrix (above).



FIG. 18. Striated muscle, longitudinal section. (670 ×) Note that the fibers do not branch or anastomose. Invisible sarcolemma envelops muscle substance. Each fiber has many flattened, slender sarcolemmal nuclei lying peripherally beneath the membrane and oriented parallel with the long axis of the fiber. In muscle disease these nuclei come to occupy a central position. Large numbers of axially disposed myofibrils are contained within the fiber. Each myofibril consists of a chain of dark contractile rods, the sarcomeres, which alternate with clear substance in forming the chain. The dark rods are perfectly aligned alongside those of other myofibrils, forming dark bands, the Q or transverse bands, which extend across the width of the muscle fiber. A fine clear line, the M line, intersects each dark band. Similarly, the clear substance of myofibrils forms light bands, the J bands, which are intersected by the thin Z line.

which are more plump than sarcolemmal nuclei and are not strictly parallel with the muscle fiber. These nuclei are particularly numerous near blood vessels and stain easily with silver impregnation methods. The enveloping connective tissue sheath is designated as the endomysial sheath

The sarcolemmal membrane functions (1) as a semipermeable membrane between the potassium-rich interior of the fiber and the sodium-and-chloride-rich exterior; (2) to transmit a stimulus so that the entire fiber contracts as a unit; (3) in conjunction with the collagenous tissue it imparts, by its elasticity, a tension to the muscle, resisting stretching and aiding contraction.

MYOFIBRILS AND STRIATIONS OF THE MUSCLE FIBER

Each muscle fiber contains a large number of axial disposed myofibrils. These are closely packed and separated by minute amounts of sarcoplasm. In cross section, the myofibrils are grouped into a number of bundles producing a maplike configurati— *Cohnheim's areas or fields.*

4

Histology of Skeletal Muscle[1]

The skeletal muscles are composed of a ge number of muscle fibers which vary ely in size from one muscle to another. average fiber diameter in most muscles is to 50 microns but may be thicker in the gest muscles and thinner in small muscles. e fiber length ranges from a few to over millimeters, varying widely within a single sciculus Fibers are joined end to end by domysial connective tissue. They do not a h or anastomose. At the tendinous end the muscle the sarcolemma merges into a ndinous fibril which joins with fibrils of her muscle fibrils to form the tendon aponurosis.

Muscle fibers are grouped in parallel arrangement in bundles called fasciculi. The fasciculi themselves run parallel and seldom extend from one end of the muscle to the other. On transverse section each fasciculus has an angular multisided figure.

At birth, larger, more mature fibers are often seen in the center of a group of smaller fibers. In the adult all fibers are equally large. However, in the course of muscle atrophy, the infantile pattern is reproduced, all but the most central fibers becoming smaller.

The muscle fiber is a multinucleated cell which contains a large number of myofibrils embedded in a matrix of undifferentiated protoplasm all enclosed within a fine sheath, the sarcolemma.

[1] Adams, R D, Denny-Brown, D., and Pearson, M Diseases of Muscle, New York, Hoeber, 1953.



FIG 17 Cross section of skeletal muscle (× 45)

SARCOLEMMA AND SARCOLEMMAL NUCLEI

The sarcolemma is the thin elastic transparent membrane which envelops the muscle substance. It normally is invisible but becomes visible when the fiber is ruptured, whereupon the contents of the fiber retract to form a granular mass and leave an empty space within the sarcolemma. It can be vitally stained with isamin blue and by janus green.

The slender, flattened sarcolemmal nuclei lie beneath the membrane and are oriented parallel with the long axis of the fiber. They are invisible in the living fiber In stained sections they are from 1 to 3 microns in width and from 5 to 12 microns in length Each fiber has many such nuclei, numbering up to several hundred. The nuclei normally lie peripherally beneath the sarcolemma except near the tendinous insertion. In fetal life and in the course of muscle disease, the sarcolemmal nuclei may occupy a central position between the myofibrils.

Outside the sarcolemma is a closely investing layer of connective tissue having a fibrillar structure and containing fibroblastic nuclei



FIG. 18 Striated muscle, longitudinal section (670 ×) Note that the fibers do not branch or anastomose Invisible sarcolemma envelops muscle substance. Each fiber has many flattened, slender sarcolemmal nuclei lying peripherally beneath the membrane and oriented parallel with the long axis of the fiber. In muscle disease these nuclei come to occupy a central position. Large numbers of axially disposed myofibrils are contained within the fiber Each myofibril consists of a chain of dark contractile rods, the sarcomeres, which alternate with clear substance in forming the chain. The dark rods are perfectly aligned alongside those of other myofibrils, forming dark bands, the Q or transverse bands, which extend across the width of the muscle fiber. A fine clear line, the M line, intersects each dark band Similarly, the clear substance of myofibrils forms light bands, the J bands, which are intersected by the thin Z line.

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SARCOPLASM

This is the undifferentiated protoplasm, an lhesive substance, which fills the spaces beveen the myofibrils and the sarcolemmal uclei The most constantly active muscles, g., respiratory, possess the greatest amount sarcoplasm, muscles that contract quickly nd fatigue easily contain the least amount. Granules of various sizes occupy this subtance and are refractile, soluble in alcohol and ether, and stain with Sudan, Scharlach R. osmic acid and Nile blue Their staining reactions lead to their designation as liposomes. They disappear rapidly after death and in certain fixing agents Some fibers are laden with fatty granules, others are entirely free of them. Glycogen is present in granular form throughout the fiber. The specific proteins of sarcoplasm are difficult to localize.

SUPPORTING TISSUES OF MUSCLE

Muscle fibers are united by connective tissue sheaths which surround each individual fiber and groups of fibers. Thus, muscle fibers are grouped into primary bundles, each containing up to several hundred fibers. The bundles are organized into one of several patterns. A muscle designed to pull in one direction is composed of a parallel arrangement of bundles and is fusiform in shape. When muscle pull is exerted in more than one direction, the fibers and the bundles have a unipennate or bipennate arrangement.

The outermost connective tissue enveloping the entire muscle is called the *epimysium.* This contains collagenous, reticular and elastic fibers, blood vessels, lymphatics and nerves. From the epimysium connective tissue extensions, the *perimysium,* penetrate the spaces between the muscle bundles Finally, from the perimysium, a delicate network of fibrous tissue, the *endomysium,* enters the muscle bundle and surrounds each fiber with capillaries, nerve filaments, fibroblasts and wandering cells. The collagenous fibril bundles wind in spirals around individual muscle fibers. The course of these fibrils varies according to the degree of contraction or stretch of the muscle. In the contracted state they are almost at right angles to the muscle fiber and close together, while in the extended position they are nearly parallel with the fibers. This arrangement permits easy displacement of muscle fibers and offers increasing resistance to stretching.

5

Physiology of Bone

COMPOSITION OF BONE

Bone is composed of approximately 33 per cent organic (protein) and 67 per cent inorganic (calcium, phosphorus, magnesium, etc.) substances. The matrix is formed by collagen fibers which are held together by mucopolysaccharide substances in the interfibrillary spaces. Calcification of these collagen fibers occurs by deposit of crystals of tricalcium phosphate in the form of a hydroxylapatite. Magnesium hydroxide is absorbed upon the crystals. The molarity ratio of calcium to phosphorus is 1:0.6.

FUNCTION OF BONE UNITS

The *periosteum* forms are enveloping layer for bone and a supportive structure through which nerves and blood vessels reach the bone. The outer or fibrous layer is supportive. The inner or cambium layer supplies bone cells for appositional cortical bone growth.

The bone cells are osteocytes, osteoblasts and osteoclasts. They are differentiated from primitive mesenchyme and may transform into one another.

The *osteoblasts* line up in rows wherever there is active bone formation. They form osteoid tissue, and produce alkaline phosphatase which promotes calcification of the osteoid tissue.

The *osteoclasts* absorb bone.

The *osteocyte* is the heart of a bone unit. By its tiny fibrils which extend through tiny canaliculi it communicates with surrounding osteocytes. It maintains the metabolism of its unit of bone. When it dies, the intercellular substance about it will resorb.

MINERALS[1]

Because bone is constantly being formed and destroyed, its maintenance depends on an adequate supply of these materials and the ability to utilize them. The presence of calcium in the blood stream is essential to life. It is necessary for (1) osteogenesis, (2) cell and capillary wall normal permeability, (3) blood clotting, (4) nerve excitability, (5) muscle irritability and (6) heart muscle action. Calcium and phosphorus comprise 85 per cent of the mineral content of bone. Less important are carbonates and magnesium. Calcium and phosphorus are ingredients of the diet and are excreted in the feces and the urine.

The fate of these minerals depends upon:

1. *The Form in Which They Exist in Food.* Human milk contains the more soluble and easily absorbed form of calcium lactose. Calcium in green vegetables exists as insoluble oxalates and therefore is not an abundant source.

2. *Food Substances Affecting Solubility and Absorption.* Excessive intake of phosphates in relation to calcium or, conversely, excessive intake of calcium in relation to phosphates, encourages precipitation of and excretion of insoluble phosphates. Excessive ingestion of fatty acids causes formation of insoluble calcium soaps. Excessive carbonates will precipitate insoluble calcium carbonate. The calcium and the phosphorus thus precipitated become unavailable for absorption.

3. *Condition of the Digestive Tract.* Gastric acidity is necessary for solubility of calcium and phosphorus. Pancreatic enzymes digest fats completely so that fatty acids cannot form insoluble calcium soaps. Any disease process of the small intestine interferes with absorption of minerals.

4. *Vitamin D.* This vitamin is mainly necessar[illegible]

phor[illegible]

5. [illegible]

thyroids controls the serum level of calcium and phosphorus by effecting resorption of bone

[1] Stein, I., Stein, R. O., and Beller, M. L.: Living Bone in Health and Disease, Philadelphia, Lippincott, 1955.

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6 *Condition of the Kidneys* Phosphorus
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An adult after the age of bone growth will
e in normal calcium and phosphorus balance,
e, he will excrete the same amount of min-
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he intake will be greater than the excretion.
When the individual is in negative balance,
he output exceeding the intake, a diseased
tate exists

Phosphorus

Phosphorus exists as a completely ionized inorganic phosphate in the blood serum. Of the mineral in the body 80 per cent resides the skeleton where it is combined with cal- as a complex apatite or triple phosphate. Phosphate in bone consists of two types: (1) a *labile fraction*, which is in equilibrium with phosphate ions in the blood, and (2) a *stable fraction*, which is fixed in the skeleton

The minimum daily requirement in the normal adult is 0 88 Gm., and slightly larger for growing children and pregnant women. Food sources are mainly from milk, and lesser amounts from meat, eggs, cheese, nuts and whole cereal. White flour and rice have a small content. Phosphorus exists in food in both inorganic and organic forms.

Absorption takes place from the small intestine in the form of soluble inorganic phosphate. The complex organic phosphorus-containing nucleoproteins must be broken down by enzymes of the pancreas and the succus entericus. The resultant phosphoric acid is hydrolyzed by phosphatase, which occurs in abundance in the intestinal wall. Phosphorus is then transported across the cell membrane in combination with calcium. Absorption is dependent upon vitamin D.

An excess of ingested calcium encourages formation of insoluble phosphates, thereby interfering with absorption of phosphorus. Secondarily, the lowered phosphorus in the blood stream will cause its removal from the bone. Conversely, if ingested calcium is inadequate, a relative excess of phosphates exists, and again insoluble phosphates will be deposited. Therefore, a proper balance of these minerals is necessary for solubility, and vitamin D is required for absorption before calcium and phosphorus can be made available to the body.

The normal level of blood serum phosphorus, which exists as ionized inorganic phosphate, is 3 to 4 mg. per 100 cc. in the adult, and 5 to 6 mg. in the infant.

Excretion takes place principally in the urine as monosodium (acid) or disodium (alkaline) phosphates, and in lesser amounts as salts of potassium, ammonium, calcium and magnesium. Ninety per cent of excreted phosphorus is in the inorganic form.

An increased excretion of phosphorus in the urine occurs with an increased dietary intake as a high-protein diet, protein catabolism as after energetic exercise, gout and hyperparathyroidism.

A low urinary excretion takes place in low-phosphorus rickets and osteomalacia, renal glomerular disease (associated with hyperphosphatemia), pregnancy and hypoparathyroidism.

Calcium

Calcium is indispensable to life. About 99 per cent of body calcium is stored in the skeleton as phosphates, carbonates and hydroxides.

The normal daily requirement for a normal adult of 70 Kg. is 0 65 Gm. and 1 Gm. for growing children and pregnant women. Larger amounts are especially required during the last trimester of pregnancy and during lactation. Only milk and milk products provide satisfactory dietary sources of calcium. Certain green vegetables, such as spinach, although high in calcium content, are not suitable sources, because their oxalic acid content forms insoluble compounds with calcium.

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and by encouraging urinary excretion of phosphorus.

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Vitamin C is necessary for the production of intercellular cement substances. Therefore, a deficiency results in a reduced capacity of fibroblasts, osteoblasts and odontoblasts to form collagen, osteoid and dentin. Matrix formation is poor. Insufficient collagen formation is reflected in slow organization of hematomas and delayed wound healing. Failure to produce osteoid interferes with endochondral ossification. At the epiphyseal plate, calcified cartilage piles up but is not replaced by osteoid and bone. The bone which does form is slender and fragile. Blood vessels, particularly capillaries, are fragile and easily ruptured, since multiple capillary hemorrhages are a feature of vitamin C deficiency.

Formation and growth and calcification of cartilage is not dependent upon ascorbic acid.

Vitamin C deficiency, or scurvy, is discussed in the section on Metabolic Diseases.

Vitamin D

Vitamin D is a fat-soluble vitamin which exists together in nature with vitamin A. Ergosterol, the synthetic counterpart of vitamin D, is formed by the action of sunlight or ultraviolet on the skin.

Vitamin D aids absorption of calcium from the intestine and is necessary for deposition of the mineral in bone. Its lack results in failure of the process of calcification of cartilage and osteoid tissue necessary to bone growth and produces rickets. On the other hand, an excess of vitamin D effects, perhaps by stimulating the parathyroids, solution of calcium from the bone, elevation of serum calcium levels, metastatic calcium deposits and increased urinary excretion with formation of calcium phosphate casts and stones. Albright believes that this chain of events is secondary to increased phosphorus excretion caused directly by vitamin D. Ingestion of excessive amounts of vitamin D, for example in treating vitamin-D-resistant rickets, must be controlled by laboratory tests A very positive urine Sulkowitch reaction and the finding of calcium phosphate casts indicates a hypercalcemia. A serum calcium value of 13 mg. or above indicates that the dose of the vitamin should be reduced.

Vitamin D deficiency or rickets is discussed in the section on Metabolic Diseases.

Vitamin E

Vitamin E, the tocopherols, because of their effect on sexual development, naturally influences production of sex hormones. Consequently, deficiency of this vitamin may be related to growth disturbances. The vitamin also inhibits formation of collagenous connective tissue. Its efficacy in the treatment of collagenous diseases is questionable.

GLANDS OF INTERNAL SECRETION

Parathyroids

These are small, brownish-red bodies measuring about 5 mm. by 3 mm. by 1 mm.; they resemble flattened oval disks. They are located in the posterior capsule of the thyroid gland and usually consist of 2 superior and 2 inferior glands, although their number may be as high as 10. Aberrant parathyroid tissue may be found in the neck and the upper mediastinum. The hormone, parathormone, has the following functions:

1. Maintains blood calcium level by resorption of bone and by controlling urinary excretion.
2. Lowers serum phosphorus level by inhibiting tubular reabsorption of excreted phosphorus.
3. Increases diuresis of calcium and phosphorus.
4. Stimulates osteoclasis.
5. Directly effects dissolution of bone.
6. Increases solubility of calcium and phosphorus.
7. Inhibits the calcifying effect of vitamin D.

Symptoms of hypoparathyroidism most often follow accidental removal of the glands during operation for a diseased thyroid. Hyperparathyroid symptoms are due to hyperplasia, or neoplasm. (See section on Metabolic Diseases.)

Gonads

Estrogens and androgens are present in both male and female. In the adult female, androgen values are almost as abundant as in the male. Although the main functions of these hormones are related to development of the genitalia and secondary sex characteristics, they also have certain important influences upon the skeletal system.

VITAMINS

Vitamin A

Vitamin A is found in greatest abundance in animal and fish liver, egg yolk, butter and cream. Vegetables with yellow or green coloring contain provitamin A which is converted to vitamin A in the animal. Absorption of vitamin A from the gastro-intestinal tract is dependent upon and parallels fat absorption. Therefore, conditions which will impair fat absorption will decrease vitamin A absorption. These conditions include idiopathic steatorrhea, pancreatic cyst, obstruction of the biliary passages, and hepatic disease. These diseases display decreased plasma vitamin A values below the normal 33.2 mcg. per 100 cc. When vitamin A is dissolved in an aqueous medium (water-propylene glycol mixture), it is absorbed readily, regardless of the degree of fat absorption.

Vitamin A deficiency is characterized by retardation of skeletal growth and maturation, epithelial hyperkeratosis and impaired ability to see in the dark.

Hypervitaminosis A (Chronic Vitamin A Poisoning). The normal daily requirement of vitamin A for infants and young children is from 1,500 to 2,500 I.U. When over a period of months large amounts are ingested, e.g., 100,000 to 500,000 I.U. daily, certain clinical phenomena appear. Tender, painful swellings develop over the course of long bones, commonly the ulna, the tibia and the metatarsals. These swellings are firmly fixed to the bone. The overlying skin is freely movable, and inflammatory signs are conspicuously absent. Limitation of movement probably is due to protective muscle spasm. The patient invariably is an infant beyond 12 months of age or a young child.

X-ray investigation reveals that the swellings correspond to areas of periosteal ossification. Usually several bones are involved, most frequently the ulna and the metatarsals. After vitamin A is discontinued, the hyperostosis shrinks over a period of several months, and the bone resumes its normal appearance.

The plasma vitamin A level exceeds the normal upper limit of 33.2 mcg. (150 I.U.).

The condition must be differentiated from infantile cortical hyperostoses, because both conditions are featured by similar tender swellings of ossification. Infantile cortical hyperostosis develops before the 4th month of infancy, whereas hypervitaminosis A occurs after the 12th month. The former always affects the face and the jaws, while the latter does not involve these structures. Fever is an invariable accompaniment of infantile cortical hyperostosis, whereas the infant with hypervitaminosis A is afebrile. The plasma vitamin A in infantile cortical hyperostosis is normal, while in vitamin A excess it is high.

Treatment requires discontinuing the vitamin. The symptoms and the findings subside within a week.

Vitamin B

Certain clinical states produced by vitamin B deficiency are of interest to the orthopaedic surgeon. The vitamin is necessary to the integrity of the central and the peripheral nervous systems. Vitamin B_1 (thiamine) and B_{12} deficiency is a common cause of peripheral neuritis.

Vitamin B deficiency, particularly of riboflavin, during pregnancy may produce congenital deformities. Experimentally, the defects can be produced almost invariably in animals by withholding riboflavin. The anomaly most likely is obtained in that limb bud in the most active state of development at the time of the vitamin deficiency.

Vitamin C

Vitamin C is found in citrus fruits, tomatoes and certain vegetables, as potatoes. Heat, sunlight and oxidation processes easily destroy it. A synthetic form of ascorbic acid can be administered to assure adequate intake.

When vitamin C intake is restricted, the plasma level may be maintained while the tissues are being depleted. Therefore, single plasma vitamin C level determinations are of no value. Multiple determinations are necessary. When vitamin C is being excreted in detectable amounts, the tissues are regarded as satisfied, and the excreted ascorbic acid represents the excess. The daily recommended intake varies from 30 mg. per day for infants to 100 mg. for adults. Under periods of stress and fever, daily requirements are greater.

Estrogen. This hormone is produced by the ovarian follicle. Secretion of estrogen is controlled by pituitary secretion of gonadotropin. Estrogen stimulates the formation of bone by its protein anabolic effect. The hormone accelerates longitudinal growth but also speeds up skeletal maturation so that epiphyseodiaphyseal union occurs prematurely. Therefore, the hormone when administered will cause an initial spurt in growth, but such growth is halted early, and the over-all stature will be less than normal.

In primary hypogonadism, a *deficiency of estrogenic hormone permits the epiphyseal plates to remain open for years beyond the usual time of closure*, active longitudinal growth continuing. The characteristic picture of a long slender, poorly muscled, hypogonadal individual is produced. The eunuch is the rarely seen extreme form of this condition.

In contrast, an excess of pituitary growth hormone causes rapid growth rate without influencing maturation and epiphyseal closure.

Therefore the skeletal effects of gonadal insufficiency include excessive long-bone growth, delayed epiphyseal plate closure, thinned trabeculations and thinned cortices.

Androgen. In embryologic development the testes and the adrenal cortices arise from adjacent structures. Thus is explained the production of similarly acting hormones from these two sites. Androgens are responsible for the development of masculinizing characteristics, e.g., hirsutism, well-developed muscles, deep voice, etc. The testis is under the control of gonadal-stimulating hormones from the anterior pituitary. Pituitary adrenocorticotropic hormone controls the adrenal cortex. Adrenal and testicular androgens are metabolized and secreted in the urine as 17-ketosteroids.

Human androgen has a protein anabolic effect, which is exerted upon muscles, skeleton, sex organs and other structures. When androgen is administered it causes retention of nitrogen, sodium, potassium and chlorides. The protein anabolic effect is reflected in increased size of muscles, stimulation of growth, and premature epiphyseal closure. When testosterone is administered to a child, an early spurt of growth occurs, but skeletal maturation is accelerated so that epiphyseal fusion is premature, and the individual possesses a shortened stature.

Uses of Sex Hormones. The main uses of androgen and estrogen are in situations requiring the protein anabolic effect for osteoblastic activity. The treatment is effective in generalized osteoporosis, particularly of the menopausal type. Slipped upper femoral epiphysis often occurs in the hypogonadal type of individual. The epiphyseal separation takes place through an area in the epiphyseal plate weakened by excessive proliferation of cartilage cells. Sex hormones can be used to accelerate epiphyseal fusion.

Administration of sex hormones should not be indiscriminate. Estrogen can stimulate the formation of breast tumors and often causes uncontrollable menorrhagia. Testosterone can inhibit pituitary secretion and thereby *suppress adrenal cortical function. This explains* reduction of urinary 17-ketosteroids occurring during androgen therapy.

Pituitary

The glandular cells of the pars anterior of the pituitary are composed of equal numbers of *chromaphobes* and *chromaphils.* The chromaphobes are smaller and take the stain poorly. The chromaphils, by their staining reactions to hematoxylin and eosin, are composed mainly of centrally located eosinophils *(pink-staining) and a lesser amount of peripherally* situated basophils (blue-staining). It is generally accepted that cells can transform from chromaphobes to chromaphils and vice versa. The clinical pictures produced by an increase of either acidophils or basophils indicate that these cells are the source of hormones. Three hormones are produced: (1) growth hormone, (2) lactogenic hormone and (3) tropic hormones. The tropic hormones are of several types, corresponding to their effects upon other glands: (1) thyrotropin, (2) adrenocorticotropin, (3) gonadotropin *and (4)* lesser understood hormones affecting in some way the parathyroids and the pancreas.

Growth Hormone. In the epiphyseal growth plate, proliferation and hypertrophy of chondrocytes are controlled by growth hormone secreted by the acidophilic cells of the pituitary gland. Complete deprivation of growth hormone, e.g., after removal of the pituitary

ossific foci until quite late. Ossification may be delayed in the spine. Characteristically, the anterior surface of vertebral bodies remain persistently notched until adulthood. The epiphyseal growth plates may remain open for many years.

Treatment. Thyroid extract is specific.

ADRENALS[5]

The adrenals play a prominent role in new bone formation, endochondral ossification and skeletal maturation. Although the severe type of adrenal hyperfunction is rare, the mild type with a paucity of symptoms is now recognized as very common and can be identified by appropriate laboratory procedures. An investigation into the causation of osteoporosis, frequent fractures and delay in union after a fracture should include blood and urinary 17-ketosteroid and 17-hydroxycorticoid level determinations. The following section deals only with those adrenal conditions which affect the musculoskeletal structures.

Hormones

According to present concepts, the normal human adrenal cortex secretes three groups of hormones.

Glucocorticoids. These inhibit protein anabolism (or further catabolism), increase gluconeogenesis (by breakdown of protein), decrease pituitary secretion of ACTH and in large amounts increase the urinary excretion of potassium and tubular reabsorption of sodium. These hormones are the probable precursors of 17-hydroxycorticoids found in the blood and the urine. Hydrocortisone is the most important glucocorticoid in man.

Androgens. These stimulate protein anabolism, accelerate endochondral ossification and skeletal maturation (early closure of epiphyseal lines) and in excessive amounts cause virilization. Androgens are the probable precursors of 17-ketosteroids secreted in the urine.

Mineralocorticoids. An electrolyte-regulating substance, aldosterone, increases tubular resorption of sodium, increases excretion of potassium and influences the distribution of sodium and potassium throughout the body.

[5] Thorn, G W., Goldfien, A., and Nelson, D. H.: The treatment of adrenal dysfunction, M. Clin. North America 40 1261, 1956

DETERMINATION OF CORTICAL FUNCTION

The first two groups of hormones are secreted in appreciable quantity only in response to elaboration of ACTH by the pituitary. The determination of 17-ketosteroids and 17-hydroxycorticoids in the urine and the blood can be used to assess adrenal function. A quantitative estimation of adrenal cortical reserve and responsiveness can be measured by administering ACTH and noting changes in the blood and the urine steroids. When hyperactivity of the adrenal cortex is due primarily to pathology within the adrenal itself, removal of pituitary stimulation will have no great effect on the 17-ketosteroids and 17-hydroxycorticoids. These substances will remain at a high level in spite of suppression of endogenous ACTH activity by injections of fluorohydrocortisone. On the other hand, if adrenal hyperactivity is due to pituitary stimulation, suppression of ACTH activity results in a marked drop of steroid levels.

Clinical Syndromes of Adrenal Hyperfunction

Hypersecretion of adrenal steroids is due to pituitary or adrenal tumors, benign or malignant, or hyperplasia. The syndrome produced depends on the relative amounts of these hormones and may even reflect an excess of a single hormone. Thus, excessive glucosteroid secretion will produce loss of tissue protein as manifest by muscle atrophy, osteoporosis and striae. On the other hand, excessive androgen excretion, while causing hirsutism, will tend to counteract protein catabolism so that muscles are well developed, bony structure is adequate, and striae are absent. The following syndromes are of orthopaedic importance.

Cushing's Disease. A basophilic adenoma or hyperplasia of the basophilic components of the anterior lobe of the pituitary gland produce the classic picture of hypertension, diabetes, osteoporosis, abdominal striae, moon face, buffalo hump, truncal obesity and hirsutism. Treatment consists of high-voltage irradiation of the gland, hypophysectomy or a combination of these procedures.

Cushing's Syndrome. In the vast majority of instances, excessive glucocorticoid production is due to pathology in the adrenal cortex, namely, hyperplasia, adrenal carcinoma and a benign adenoma. Preoperative diagnosis to determine the type of lesion is extremely diffi-

is characterized by exaggeration of ossification unevenly distributed throughout the body.

The osteoblasts are stimulated to overfunction beneath the periosteum but particularly at points of stress and pull (muscle and ligament attachments) and points of compression (ends of long bones). The growth of bone is greatly exaggerated characteristically in the mandible, the malar bone, the skull (especially at the frontal area) and at the extremities of long bones Bony prominences may protrude from the inner table of the skull. The thorax is massive The vertebral bodies are enlarged. particularly anteriorly. The pituitary fossa may be enlarged because of the expanding influence of the pituitary tumor. Acromegalic arthritis resembles ordinary osteoarthritis in that the subchondral bone is thickened, and the articular cartilage is worn away at the points of greatest compression. Histologically, the greatest activity of osteoblasts is seen at the points of attachment of ligament or muscle where it seems that bone production is stimulated by the intermittent pull of these structures. In addition, gigantism has the element of excessive longitudinal growth and a more diffuse increase in size of all the bones.

Thyroid

The hormone thyroxin has some direct or indirect effect upon bone growth. Hypothyroidism during the growth period results in *stunted growth. Hyperthyroidism is associated* with generalized demineralization of the skeleton. A plausible explanation is the excessive protein catabolism of a hypermetabolic state.

Thyroid Insufficiency. Lack of thyroid hormone is due to congenital absence of thyroid tissue, or to postnatal hypofunction of the gland. Insufficient thyroid is reflected in delayed growth and maturation of cartilage and slowed endochondral and intramembranous ossification. Consequently, the effect upon the skeletal system is most pronounced during the growth period of infancy and childhood.

Cretinism. The hypothyroid newborn infant, the cretin, is normal at birth, because of prenatal influence of maternal hormone. Gradually, within the first few weeks or months, the characteristics of cretinism become manifest, i.e., anorexia, constipation, failure to gain weight, sluggishness and a hoarse cry. The face appears simian, dull, pale; the tongue protrudes; the bridge of the nose is flattened; and mental development is retarded. Dentition is delayed, and longitudinal growth is slowed, resulting in dwarfism. Fontanel closure is delayed.

Myxedema. This term is applied to the hypothyroid state developing after birth. However, when it occurs during the growth period, the effects are similar to cretinism, although of lesser degree, because thyroid hypofunction is partial. Mild symptoms are such as to escape notice and include anorexia, lassitude, easy fatigue and sluggish behavior. Nevertheless, longitudinal growth is retarded sufficiently to produce lessened height.

Laboratory Findings. The following pertinent tests are diagnostic:

1. *Low Basal Metabolism.* This is difficult to perform on infants and children and is often normal in subclinical hypothyroid states
2. *Protein-Bound Iodine* (PBI). The normal is 5.0 to 7.0 mcg. per 100 cc. In hypothyroidism, it may be reduced as much as one half normal. It is most reliable for detecting subclinical states.
3. *Radioactive Iodine Uptake.* The normal is 10 to 15 per cent of the administered dose. A cretin displays a trace of iodine uptake.
4. *Hypercholesterolemia.* This is frequently associated with hypothyroidism.
5. *Increased serum carotene* is often but not invariably found.
6. *Serum Calcium and Phosphorus Are Normal.*
7. *Serum alkaline phosphatase is reduced* reflecting decreased osteoblastic activity.

Roentgenographic Findings. In the infant, ossification of membranous bones of the skull is delayed, and fontanel closure is late. Pneumatization of sinuses is delayed and incomplete. Later, ossification of bone developing in cartilage is delayed. The ossification centers of epiphyses are late in appearing; and when they do, multiple foci of ossification develop in each epiphysis, a characteristic appearance designated as *cretinoid epiphyseal dysgenesis.*[2, 3, 4] Carpals and tarsals do not exhibit

[2] Wilkins, L: Epiphyseal Dysgenesis associated with hypothyroidism, Am. J. Dis Child 61.13, 1941.
[3] Wilkins, L, Fleischmann, W., and Block, W.: Hypothyroidism in childhood, J. Clin Endocrinol. 1 3, 1941.
[4] Quimby, E H., and McCune, D. J: Uptake of radioactive iodine by the normal and disordered thyroid gland in children, Radiology 49.201, 1947.

deficient in gonadotropic hormone. Consequently, they are underdeveloped sexually. Slipped epiphysis, which frequently occurs in the Fröhlich type of individual, is benefited by gonadotropic hormone.

ENZYMES

Normally, alkaline phosphatase occurs in greatest concentration at (1) the intestinal mucosa, (2) bone and (3) the kidney.[7] In other words, it functions principally at sites of absorption, deposition and excretion of calcium and phosphorus. In bone, it is concentrated at the main points of ossification, i.e., the epiphyseal line and the subperiosteal area. During active bone destruction, a compensatory stimulation of osteoblasts to form phosphatase in an attempt to replace the bone is reflected in an increased level of alkaline phosphatase in the blood stream. Because it is present in large concentration at points of active bone formation, the phosphatase stain method of Gomori is used to study bone formation. The normal range in adults is 0.5 to 4.0 Bodansky units/100 cc; in children, 5.0 to 14.0 units/100 cc. A marked increase as high as 135 units is found in Paget's disease. Hyperparathyroidism is associated with a moderate rise, as in active rickets. A slight to moderate rise in osteoblastic osteogenic sarcoma is proportionate to the amount of new bone formation. Diseases of the liver with jaundice typically show an elevated serum alkaline phosphatase. This is due to failure of elimination of the constantly formed enzyme. The counterpart of this enzyme is acid phosphatase, an enzyme found in various tissues but principally in the adult human prostate. The normal serum level is 0.0 to 1.0 Bodansky unit. A definite increase occurs in carcinoma of the prostate with metastases.

Phosphorylase is an enzyme which prepares glycogen for phosphatase effect on growing cartilage and possibly bone.

Pathologic Physiology of Alkaline Phosphatase. Experimental work has demonstrated that the formation of alkaline phosphatase is closely related to young fibroblasts concerned with the deposition of the fibrocollagenous framework or matrix of bone rather than with impregnation of this framework with calcium salts. By stains specific for the enzyme, the position and the concentration of alkaline phosphatase can be detected in the tissues. Thus fibroblasts in the outer layers of periosteum are lacking in enzyme, while those in the cambium layer where they are being differentiated into the osteoblastic stage contain large amounts of the enzyme. Staining identifies the enzyme as being intranuclear, intracellular and extracellular. The fibrils which are thrown out to form collagenous material, the precursor of the matrix, are intensely stained.

Staining for the enzyme is useful for studying osteoblastic activity. For example, osteoblasts and their precursors with the alkaline phosphatase can be seen about a bone transplant effecting creeping substitution. When new fibrocollagenous matrix is formed, osteoblasts can be traced to their destiny. Some osteoblasts persist as osteocytes. Others become cells within the matrix not demonstrable by ordinary hematoxylin and eosin stain, because the nucleus loses its basophilic staining but retains its staining for alkaline phosphatase. As the osteoid forms, this cell disappears, and alkaline phosphatase is no longer demonstrable. It appears that the next stage, namely calcification, is not dependent upon alkaline phosphatase.[8]

Alkaline phosphatase is closely linked with the formation of a fiberlike material whether or not these fibers form bone. It is related to young fibroblasts and the building material between the cells, both normal and pathologic. The enzyme is present in large quantities in fibrosarcoma, in polyostotic fibrous dysplasia, and the fibrous matrix of bone from Paget's disease.

Acid Phosphatase. This enzyme is capable of hydrolyzing hexosediphosphate at a pH of 5. It is found in large concentration in the prostate and in lesser amounts in the seminal vesicles, the testes, the epididymis and the spermatic duct. It appears in large amounts in the blood stream in metastatic carcinoma of the prostate, even before bone involvement is apparent on x-ray examination. The alkaline phosphate may also rise in this condition.

[7] Kabat and Furth Histochemical study of alkaline phosphatase, Am. J. Path. 17:303, 1941.

[8] McKelvey, A. M., and Mann, F. C.: Role of alkaline phosphatase in osteogenesis after transplantation of bone, Am. J. Path. 25:709, 1949.

cult. Steroid determinations are helpful. *Benign hyperplasia or adenoma usually displays a high basal level of 17-hydroxycorticoids and normal or slightly increased 17-ketosteroids. Carcinoma is characterized by markedly increased levels of both hormones.* Finally, one can demonstrate that the hyperfunction is independent of the pituitary; suppression of endogenous ACTH by administration of cortisone or fluorohydrocortisone fails to reduce materially the blood and the urine steroid level. Occasionally, roentgenographic examination by intravenous pyelograms and presacral air insufflation may indicate the location of the tumor.

Treatment depends on the intensity of symptoms. For *mild* Cushing's syndrome, pituitary irradiation is indicated. Response to x-ray is slow but results in a temporary or a permanent remission Surgical treatment is recommended for patients suspected of having an adrenal tumor as well as those with *severe* symptoms and those who fail to respond to pituitary irradiation. In the absence of an adrenal tumor, total bilateral adrenalectomy followed by hormonal replacement therapy appears to be the treatment of choice. Postoperatively, a high daily level of cortisone is maintained, then gradually reduced to less than 75 mg, when a sodium-retaining hormone is added. Testosterone must be given for marked osteoporosis and muscle atrophy. Cortisone therapy is continued indefinitely after bilateral extirpation. A unilateral adenoma is associated with atrophy of the contralateral gland. Following removal of the adenoma, cortisone and ACTH are administered until the suppressed gland has been restored.

Patients with Cushing's syndrome are susceptible to compression fractures, because of protein loss and osteoporosis. Testosterone encourages protein anabolism. Potassium chloride (6 to 9 Gm. daily), replenishes the mineral and reduces sodium retention Steroid diabetes is resistant to insulin but fortunately does not cause acidosis

Congenital Adrenal Hyperplasia. The defect in these adrenals is the inability to produce normal amounts of hydrocortisone Consequently, the pituitary secretes excessive amounts of ACTH leading to adrenal hyperplasia. Increased amounts of androgenic steroids are produced, resulting in virilization and excessive urinary excretion of 17-ketosteroids. This adrenogenital syndrome causes precocious puberty in the male and pseudohermaphroditism in the female. Growth is rapid, but premature epiphyseal closure results in short stature.

Treatment requires correcting hormonal deficiency. Cortisone or hydrocortisone will suppress ACTH production and lessen stimulation of the adrenal cortex to produce androgenic compounds. Adequacy of dosage is determined by reduction of urinary 17-ketosteroids to normal and restoration of a normal growth curve.

Relationship of the Adrenals to Stress. Stress such as that produced by trauma or infection is counteracted by hyperfunction of the adrenal cortex. Selye has observed that hypertrophy of the adrenal cortex is invariably associated with the "alarm reaction." Adequate response of the adrenal gland to injury is reflected in a drop in the eosinophilic level in the circulating blood. During eosinopenia, the patient is ill and, as improvement sets in, the eosinophil count rises. Eosinopenia persisting beyond the usual stress period, e.g., for many days after a compound fracture or surgery, suggests a complication, such as infection. If, after operation or severe injury, eosinophilia persists and the course is unsatisfactory, adrenal insufficiency is apparent and demands urgent measures. Cortisone must be administered in large doses. As the patient's condition improves, ACTH is given to stimulate adrenal function [6]

Physiologic Effects of Cortisone and Hydrocortisone. A summary of the physiologic actions of compounds E and F is described in the section on rheumatoid arthritis.

Chorionic Gonadotropin

This hormone is found in large amounts in the urine of pregnant women. It is a potent stimulator of interstitial cells of the testicle, increasing formation of male sex hormone and encouraging descent of the testicle. It also causes a spurt in growth. Pituitary dwarfs are deficient in growth hormone and usually are

[6] Nicholas, J A., and Wilson, P D · Adrenocortical response to operative procedures on bones and joints, J. Bone & Joint Surg. 35-A:559, 1953

phosphatase decreases. When ossification takes place, chondroitin sulfate rapidly disappears, and large amounts of alkaline phosphatase appear. The removal of acid-reaction chondroitin sulfate is a prerequisite for the action of osteoblasts. This may occur simply by a hydrogen ion shift to the alkaline side which depolymerizes the chain molecule. Other factors such as proteolytic enzymes must also be considered.

PHYSIOLOGY OF JOINTS

Articulations serve the purposes of motion and weight-bearing. The articular capsule, which consists of tough, fibrous and modified connective tissue, encloses with the epiphyseal cartilages a potential space filled with a hypocellular fluid of high viscosity. Articular construction allows ease of motion to be combined with stability. The viscous synovial fluid aids by its cohesive action in uniting the articular ends and forms a strong fluid film upon which the cartilaginous surfaces glide with negligible friction. The capsule and the tendons inserting into it, the ligaments and the muscular tone, impart steadiness to the joint, while cartilage by virtue of its remarkable elasticity buffers the impacts to the rigid skeleton. Cartilage under normal conditions and particularly with advancing age is subject to wear and tear to a degree unequaled by other tissue. In order to afford resistance to pressures, fragile structures (vessels and nerves) are excluded from cartilage. Therefore, its subsistence depends upon synovial fluid. Dyes placed in a joint can diffuse into the cartilage along the intercellular system of fibrils. Arterial blood does not come in contact with cartilage cells. Therefore, breakdown of glycogen takes place by anaerobic oxidation as shown by the low respiratory quotient of this tissue.[11, 12]

Synovial fluid is a dialysate of blood plasma as indicated by the distribution of electrolytes and nonelectrolytes between blood and synovial fluid, and the marked vascularity of the synovial membrane. It contains albumin and globulin through capillary permeability; it contains mucin, the origin of which is unknown. However, evidence to date indicates that it is formed by the connective tissue cells of the synovium and is carried into the joint by the plasma dialysate. The opposing forces of capillary pressure and osmotic pressure between plasma and synovial fluid determines the amount of fluid in the joint. Compared with true membranes, the synovium has a markedly greater permeability. Resorption of small molecules takes place almost entirely by the blood vascular system and only to a slight degree by the lymphatics, while the larger protein molecules are removed with difficulty and only by way of the lymphatics. The predominant cellular constituents of synovial fluid are mononuclear phagocytes. They carry particulate matter and the cellular debris of wear and tear from the joint into the lymphatics in a manner which suggests that the synovial membrane participates in reticuloendothelial activity. The maintenance of a normal amount of synovial fluid depends on all these factors. Perhaps the slightly negative intra-articular pressure, varying with motion, also plays a role in this equilibrium. The effect of the autonomic nervous system through its effect on blood vessels and cellular activity is unknown.

Deviations from normal articular physiology consist of alterations in synovial tissue and changes in intra-articular metabolism. The former leads to quantitative and qualitative disturbances of the exchange equilibrium; the latter results in deficient nutrition to cartilage. The available mucin may be reduced. Cartilage, once injured, does not regenerate but is repaired by invasion of fibrous tissue which in turn undergoes metaplasia into cartilage.

Normal synovial fluid is a relatively acellular, clear, straw-colored, viscous liquid. Its pH is similar to serum pH. Its protein content consists of albumin chiefly but also globulin and mucin. Fibrinogen is absent. Synovial fluid resembles tissue fluid in its cytology and mucin and may be merely tissue fluid.

Altered Physiology and Structure of Joints

As long as a joint fulfills its function of weight-bearing, stability and motion, its physiology and structure remain unaltered. However, it will react adversely to any *trauma*, whether this be *mechanical* or *irritant*. Mechanical factors include one severe impact to

[11] Ropes, M. W., Bennett, G. A., and Bauer, W.: The origin and nature of normal synovial fluid, J. Clin. Invest. 18:351, 1939.

[12] Bauer, W., Ropes, M. W., and Waine, H.: The physiology of articular structures, Physiol. Rev. 20:272, 1940.

Sometimes the osteoblastic phase of Paget's disease or the osteoblastic type of osteogenic sarcoma may be confused roentgenologically with metastatic carcinoma of the prostate. In Paget's disease only the alkaline phosphatase is raised considerably. In osteogenic sarcoma the alkaline phosphatase may be raised slightly to moderately. In prostatic carcinoma, both alkaline and especially the acid phosphatase are increased.

CARTILAGE PHYSIOLOGY

Cartilage, by its rubbery resiliency, functions to *reduce pressure*, and, where it covers the end of a bone, its smooth surface *minimizes the friction* effect of shearing stresses Where pressure and shearing stresses are brought to bear at skeletal junctions, cartilage is prone to form Thus a rib is joined to the sternum by a segment of cartilage. When movement between osseous structures is small and pressure constitutes the main force, the collagenous component of cartilage is increased, i e., the fibrocartilage of the intervertebral disk. When movement and shearing stresses are maximal, a synovial-lined cleft forms to separate two cartilage-covered surfaces, the amphiarthrodial joint

The matrix of cartilage is composed of a gel, a glycoprotein, which on hydrolysis yields chondroitin sulfate. The healthy state of cartilage is related in some way to a high chondroitin content. Conversely, degenerated cartilage has a low amount of chondroitin.[9, 10]

Qualities of Chondroitin Sulfate. This is a long-chain structure with a high molecular weight and is slightly viscous in solution. It is acid in reaction and is easily depolymerized on extraction with a weak alkali. In cartilage it is combined with protein.

Methods of Determination. The *macrochemical* method involves extraction by a weak alkali and analysis. The *histochemical* assay depends upon a metachromatic staining reaction and constitutes a qualitative test.

Origin. The intense staining reaction about chondrocytes suggests that these cells form chondroitin.

[9] Sylvén, Bengt: Cartilage & Chondroitin Sulfate. I. The physiological role of chondroitin sulfate in cartilage, J. Bone & Joint Surg. 29 745, 1947.

[10] ———: Cartilage & Chondroitin Sulfate. II Chondroitin sulfate and the physiological ossification of cartilage, J. Bone & Joint Surg 29.973, 1947.

Distribution. Each type of cartilage possesses a certain normal amount of chondroitin sulfate. The surface zones of articular cartilage contains a smaller amount than the deeper zones. A constant depletion of this substance takes place with advancing age.

Water Content. The intercellular matrix of cartilage constitutes a gel containing 70 per cent of water. The water content is dependent upon the chemical constitution of the chondroitin sulfate and the amounts of proteins and other electrolytes.

Nutrition of Cartilage. Cartilage has no blood vessels except for an occasional one passing through to other tissues. It gets its nutrition by diffusion through the perichondrium. Some histologists maintain that small canals form communicating channels between the chondrocytes. Injected dyestuffs are absorbed quickly by the cartilaginous matrix. Articular cartilage is nourished from underlying bone and by diffusion from synovial fluid.

Metabolism of Cartilage. This decreases gradually with advancing age because of a gradual decrease in the number of cells. Glycolysis and oxygen consumption are gradually reduced. It is entirely possible that constant destruction and reformation of chondroitin sulfate takes place throughout life. The formation of chondroitin may be related to other unknown factors possibly in connection with sulfur metabolism.

The synovial fluid contains *hyaluronic acid*, which resembles chondroitin chemically. Hyaluronidase (obtained from bacteria and in large amounts from the testes) hydrolyzes hyaluronic acid and chondroitin sulfate. Its origin from bacteria suggests the mechanism of cartilage destruction in infectious arthritis.

Regeneration of Cartilage. Formation of new cartilage takes place by metaplasia from new connective tissue. Cartilage cells develop slowly from perichondrial cells and fibroblasts. Heterotopic formation of cartilage, as in the synovium of chondromatosis, apparently needs only the nutritional supply from synovial fluid. When cartilage is cultured in vitro, it first dedifferentiates to a fibroblastic tissue with loss of chondroitin sulfate.

The Physiologic Ossification of Cartilage. During embryogenesis of cartilage, the content of chondroitin sulfate increases, and alkaline

Whenever an articular surface becomes irregular as a result of infection or rheumatoid involvement, the incongruity of the apposed surfaces causes friction and leads ultimately to degenerative arthritis.

The pain fibers are situated to a great extent in the synovial lining of a joint. Removal of the synovium generally relieves pain and halts production of synovial fluid. However, the remaining connective tissue is capable of regenerating the membrane and, if the original irritant factor is present, the symptoms will return.

Blood in a joint acts as a chemical irritant. It will provoke production of excess synovial fluid. When blood is admixed with synovial fluid, clotting is prevented. Therefore, a hemarthrosis in itself is not productive of adhesions inasmuch as a clot is a necessary prerequisite. Aspiration of bloody fluid does nothing more than relieve pain. It forms a good medium for infection, and removal of the blood reduces the intra-articular pressure, permitting further bleeding. A needle should not be introduced into such a joint for other than diagnostic purposes. Eventually, the blood will be resorbed.

PHYSIOLOGY OF SKELETAL MUSCLE

Skeletal muscle is completely under the control of its nerve of supply and contracts only in response to impulses reaching it through this nerve. It is elastic and can be stretched and contracts more vigorously when stretched.

When its nerve is cut, the muscle is paralyzed. However, it is still capable of contracting in response to artificial stimuli, including thermal (heat), chemical (application of salt crystals), mechanical (a blow), or electrical. Electrical current is most convenient for determining properties of muscle An induction coil produces faradic current by which a single, almost instantaneous, electrical shock of graded intensity or successive shocks can be produced. A muscle will also respond to direct (galvanic) current.

When a muscle is stimulated by a single induced shock, it contracts and then relaxes. A brief interval, the *latent period*, exists between the stimulus and the beginning of muscular contraction. Relaxation requires a slightly longer time than contraction. The time required for contraction varies for different muscles. It is generally prolonged where slow movement is usual, and short where rapid movement is necessary.

The strength of contraction depends upon many factors, including the condition of the muscle, the load which it must lift, its temperature, strength of the stimulus, etc. An electrical stimulus can be varied from a minimal stimulus (one that produces a barely perceptible contraction) to a maximal stimulus (one that can produce the strongest contraction of which that muscle is capable). Stimuli which are gradually increased from the minimal to the maximal will cause correspondingly increasing contractions. Electrical stimuli stronger than the maximal cannot bring about more than the maximal contraction. Further, stimuli stronger than the maximal may injure the muscle, resulting in decreasing strength of contraction. This response to stimuli applies to muscles as a whole. On the other hand, *individual muscle fibers follow the All-or-None Law*, which means that each fiber, when it responds, does so with a maximal contraction. Thus when a submaximal stimulus is applied to an entire muscle, the submaximal contraction of the muscle is brought about by contraction of only a portion of its fibers. The muscle as a whole does not obey the All-or-None Law. When we use our muscles we rarely call upon them for the greatest possible contraction; most voluntary contractions are submaximal, only a portion of the fibers being active. This mechanism makes it possible for certain movements to be carried on for prolonged periods of time before fatigue sets in. With each contraction a different set of muscle fibers are brought into play, enabling other fibers in the interval to regain their strength for the next contraction. The continuous functioning of postural muscles throughout the day probably operates and maintains their efficiency in this manner.

The response of muscle varies with temperature. At 0° C. muscle will not contract; accordingly, cold-blooded animals at this temperature are immobile. As the temperature rises, the contractions at first are slow and weak, then become rapid and vigorous. When the temperature rises above a maximal point,

the joint or a series of repeated impacts. When a deformity about a joint throws abnormally increased pressures upon one side of the joint and less on the other, this constitutes repeated trauma. The synovium becomes hyperemic and swollen, and an outpouring of synovial fluid occurs, seemingly to provide material for cushioning the impacts or for better lubrication. Repeated pressures upon the cartilage cause it to undergo fibrillation, degeneration and separation, being extruded into the joint as a free cartilaginous body. This in turn causes further damage to the joint surfaces. Eventually, the cartilaginous surface is destroyed, and the underlying cortical bone is exposed to the pressures. Bone has even less resistance to trauma and breaks down. The bone trabeculae become compressed and concentrated, the area being visible as apparent bony sclerosis Actually, bone formation is at a minimum.

Continued repeated inflammations and attempts at healing by granulation tissue results in production of fibrous tissue which thickens the capsule. The resulting joint is irregular and mechanically deficient. Motion produces friction and further synovitis, effusion and surface destruction. Gradually, the secreting power of the synovium is diminished, the joint surfaces are approximated, and motion is lessened or lost. This is the picture of degenerative arthritis. Fusion across the joint does not take place. The instability of the joint results in pulling strains of the capsule at the site of its attachment to the margins of the articulating bones The constant traction on the periosteum elevates it, and the resulting periosteal ossification produces spurs. Anything which interferes with the accurate mechanics of the articulation will result in degenerative arthritis. A foreign body, as shrapnel, or a loose osseous or cartilaginous body, such as a loose torn semilunar cartilage or the joint mice in chondromatosis will cause the joint to deteriorate.

An irritant substance will cause the synovium to react by hyperemia, swelling and increased outpouring of synovial fluid. Such irritants are *toxic,* such as in toxemias of certain infectious diseases (e.g., typhoid fever); *chemical,* such as sodium urate deposits of gout, and *bacterial,* such as gonococcus infection. Toxic and chemical substances as a rule cause no destruction of joint surfaces, the articulation returning to normal once the irritative factor has been removed. Bacteria, on the other hand, because of their osteolytic ferments produce disintegration which is proportionate to the type of organism and the time of exposure. The streptococcus is decidedly virulent and destroys the synovium and the cartilage, the invader reaching the underlying bone. The erosion into blood vessels and the speed of involvement which does not permit of an adequate defense results in a bloody serosanguineous exudate. The staphylococcus is likewise destructive and dissolves the articular cartilage, but the defense allows outpouring of purulent exudate. The gonococcus is a little less destructive. At any rate, these suppurative infections, after dissolving the surface tissues, excite the synovium to respond by production of granulation tissue and the formation of adhesions which bridge the joint and eventually may ossify. The more insidious type of infection, e g., tuberculosis, provokes a slower synovial response. The joint fluid is increased but is usually clear. The synovium is moderately or slightly hyperemic but is not edematous. Granulation tissue and fibrous proliferation cause the synovium to become thickened and adhesions to bridge the joint. Instead of being dissolved the cartilage is undermined by ingrowth of pannus beneath it or by extension of infection from the joint below. The fibrous ankylosis may become ossified.

Rheumatoid arthritis deserves special mention. Although the cause is unknown, the pathology suggests a low-grade infection or response to a toxic substance. Within the hyperemic, edematous synovium, the cells peculiar to low-grade infection, namely the lymphocytes and plasma cells, are found in localized collections, particularly about blood vessels Granulation tissue reaction results in fibrous tissue replacement. At the periphery of the joint, the granulation tissue forms abundantly and extends across the articulating surface, destroying the surface cartilage as it advances The synovial tissue becomes hypertrophied and villous but eventually is destroyed, and fibrous tissue, which later ossifies, extends across the joint.

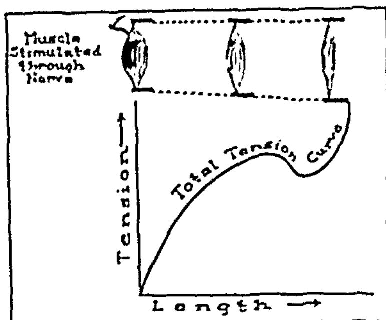


FIG. 20. At the left side of the curve, the muscle is the shortest length at which it will produce any tension when stimulated with a maximal tetanus. With the muscle tested at greater and greater lengths, the tension developed on maximal tetanus increases. *The curve then flattens off*, and the tension decreased before a final steep rise. This is called the "total tension curve." Its peculiar S shape will be better understood by observing its two components, the passive-stretch curve and the contractile-force curve (Figs. 21 and 22). (Bechtol, C. O.: Muscle physiology, Am. Acad. Orthop. Surgeons, Lect. 5:181)

than when comparable contractions are voluntary. It is generally accepted that tonus is produced by intermittent contractions of *different fibers in the muscle, which permits* continuous activity of the whole muscle, e.g., the maintenance of postural tone. The *shortening* of a muscle in tone does not amount to more than one tenth of the possible maximum, so that only 1 fiber in 10 would need to contract at any one time. Section of the motor nerve *abolishes tone.*

Prolonged and rapidly repeated muscular contraction causes fatigue. Onset of fatigue is brought on sooner by reduction of blood supply to the muscle, because substances which cause the sensation of fatigue are not readily removed by the blood stream.

A muscle which is not used undergoes atrophy. Conversely, increase in size and strength follows exercise. Activity increases the glycogen storage in muscle. When active exercise is contraindicated or impossible, massage and passive movement will reduce the degree of atrophy.

Ordinary exercise does *not increase the* amount of nitrogenous material excreted in the urine, whereas excessive activity may do so. This suggests that muscles are forced to use their own *protein material as a source of* energy. Atrophy *often follows excessive use.* Therefore, for competitive athletics a gradual build-up period to increase its glycogen capacity is desirable.

MUSCLE TENSION[13]

From the standpoint of orthopaedics, the most important property of muscle is its ability to develop tension. The force exerted upon a part is dependent upon the effective contractile power developed within the muscle acting upon that part. The degree of muscle tension is *directly related to the length of the* muscle at the time it contracts. For each *muscle there is a definite length at which* contractile power is strongest and most efficient. Determination of this length is important in surgery involving lengthening, shortening, or transplanting of muscles.

Two factors are effective in developing tension within a muscle. The first is the *contractile force*. The second is the *passive stretch* or elastic rebound resistance offered by the *sarcolemma sheaths and fibrous tissue.*[14] These can be determined for each muscle in the laboratory and plotted as curves on a graph.

THE BLIX CURVE[14]

A muscle is attached to an isometric strain gauge which will record the tension developed *by the muscle without allowing* it to shorten. With the muscle held at different lengths, it is

[13] *Bechtol, C. O.*: Muscle Physiology, Instructional Course Lectures, Am. Acad. Orth. Surg. 5:181, J. W. Edwards, Pub., 1948

[14] *Blix, M.*: Die Lange and die Spannung des Muskels, Skandin. Arch. f. Physiol. 5:150, 1894.

a state of permanent shortening called rigor ensues, and the muscle is considered as dead.

Two types of contraction are recognized. An *isotonic contraction* allows the muscle to shorten when exerted against a light movable load. An *isometric contraction* acts against an immovable object, thereby preventing shortening of muscle length. When no load is applied to the muscle the amount of shortening is at a maximum. As the load is increased, the amount of shortening becomes less, but the amount of work performed becomes greater, since work is load times the distance through which it is moved There is an optimum load with which an individual muscle can perform the greatest amount of work, and if heavier loads than this are applied, the amount of work decreases until it becomes zero, at which point the load becomes too great for the muscle to move (isometric contraction). Therefore, there is an optimum load for each muscle at which that muscle is most efficient

When a muscle is stimulated repeatedly at intervals so short that recovery is incomplete, certain phenomena will appear. The first few contractions increase in strength. This period corresponds to the "warming up" process of the athlete. Muscular contraction produces lactic acid, carbon dioxide and other by-products. The first effect of these metabolic by-products is to increase the irritability of the muscle, benefiting the first few contractions. Then the contractions begin to decrease in strength, and finally relaxation becomes so slow that the next contraction may start before relaxation is complete. Decreasing strength of contraction indicates fatigue. Failure of the muscle to relax completely is designated as *contracture*. However, this term should be reserved for muscles which are permanently contracted rather than the temporary shortening of fatigue.

The source of energy for muscular contraction is not definitely established Oxidation of dextrose or similar compounds can provide the energy, but whether other compounds such as fat can be used is questionable These oxidative processes do not provide the immediate source of energy for the contraction, but rather they supply the energy for the chemical "fuel" which is stored for instantaneous use in case a contraction is called for. When a muscle is stimulated a compound, phosphagen, composed of creatine and phosphoric acid, breaks down and liberates energy for contraction. Another compound of dextrose, probably glycogen, breaks down into lactic acid after contraction has been completed, the energy thus released being utilized to resynthesize phosphagen. Later oxidation rebuilds the lactic acid into the dextrose compound. Thus it is apparent that the oxidative processes are necessary for the formation of the chemical compound held in readiness for the contraction. Muscles can contract for some time in the absence of oxygen. The amount of work performed in a short time, such as the short burst of strenuous work of the athlete running the 50-yard dash, depends almost exclusively upon the breakdown of phosphagen and the formation of lactic acid. At the end of the run an "oxygen debt" has been acquired which is repaid by rapid breathing, thereby resynthesizing the phosphagen and lactic acid precursor.

A contracting muscle shortens in length and increases in breadth without change in volume. This activity is accompanied by a wave of electrical negativity, and formation of creatine, phosphoric acid, lactic acid and carbon dioxide Carbon dioxide is completely, and the other substances partially, removed from the muscle by the blood stream.

Repeated frequent single electrical induction shocks applied to a muscle will not allow the muscle to relax between shocks. The muscle remains in a state of contraction until it becomes fatigued. Continuous contraction is called "tetanus." If the stimuli are spaced far enough apart to allow a small amount of relaxation between stimuli, an incomplete tetanus is present. In this state the muscle shortening is greater than it would be from a single stimulus. It is probable that a voluntary muscular contraction is tetanic in nature. The multiple impulses reaching a muscle during a voluntary contraction can be measured in the number of electrical variations in the nerve with a sensitive galvanometer; they vary from 40 to 100 per second.

Muscles in a sustained state of partial contraction are said to possess *tonus*, the result of involuntary contraction. Muscles in tonus exhibit a slower rate of oxygen consumption

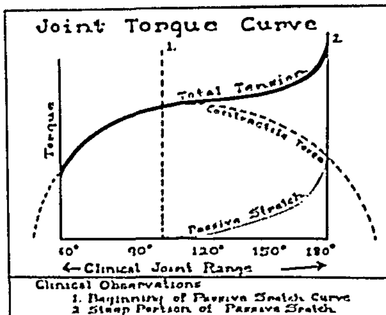


Fig. 23. The force developed by the muscle through the varying lever arm of the joint should properly be called a joint-torque curve. The extremes of the contractile-force curve are usually beyond the clinical range of the joint. The extremes of the total tension curve are considerably flattened by the compensating action of the joint lever arm. Clinically, the two important observations are: (1) the beginning of the passive-stretch curve usually corresponds with the greatest contractile force, and (2) the steep portion of the passive-stretch curve corresponds to the end of the range of the muscle and a comparatively weaker portion of the contractile-force curve. (Bechtol, C. O.: Muscle physiology, Am. Acad. Orthop. Surgeons, Lect. 5:181)

called the *passive-stretch curve* for the muscle.

Finally, by subtracting the passive stretch curve from the total-tension curve, a third curve can be drawn into the diagram which represents the contraction force and is designated the *contractile-force curve*. The contractile-force curve is highest in the center of its range and falls off markedly toward each end. This central peak of maximal contractile power corresponds to a definite length in the muscle at which passive resistance to stretch begins. Deviations from this optimum length in either direction will reduce the contractile force of the muscle. When the muscle is tested at greater than optimum length, although its total tension may be higher, a greater part of its tension is exerted by passive stretch. The latter acts as a spring which must be overcome by antagonist muscles.

The Blix curve may be demonstrated clinically by examining one's own fingers. With the wrist and the fingers in full flexion, thereby shortening the flexor muscles to their fullest extent, very little flexor power is obtainable, and no passive stretch is demonstrable at this length. With the wrist in line with the forearm, and each finger placed at 40° of flexion, flexor power is considerable. At this tendon length, the beginning of the passive-stretch curve can be detected. If the fingers and the wrist are fully extended, a marked amount of passive stretch is demonstrated. (This resistance to extension is not merely due to capsules and ligaments of the joints, since, if the tendons are relaxed by bringing the wrist back in line with the forearm, considerable hyperextension of the finger joints can be obtained.) If the contractile force is tested with the wrist and the fingers in full extension, it will be found to be weak. In this position the total tension is due largely to passive stretch.

In normal muscle the beginning of the passive-stretch curve corresponds closely to the greatest height of the contractile-force curve. This corresponds to the length that the muscle assumes when it hangs free and is designated as the true *resting length* of muscle. Passive stretch in the whole muscle is due largely to elasticity of the connective tissue and the sarcolemmal coverings of muscle fibers.

When a Blix curve is run with a fatigued muscle, the total tension line appears as a straight line, because only a small amount of tension can be developed. A muscle that is fibrosed and shortened will develop an earlier passive-stretch curve so that the contractile-force peak appears earlier and therefore is of lesser force than normal.

CLINICAL APPLICATIONS

Tendon Transfers

At the time of operation, the tendon of insertion of the muscle is divided, and the be-

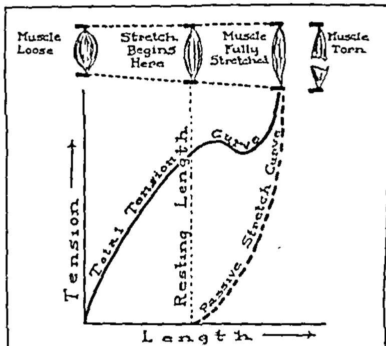


Fig. 21. The total tension curve of Fig. 20 is traced into this figure for the sake of comparison. If the muscle is re-tested over the same range, this time observing not the tension developed under a maximal tetanic stimulation, but merely the passive stretch of the muscle when [illegible] the muscle hangs loosely between the supports. At about the center of the curve, it begins to exert some pull after the manner of an elastic band. Owing to its elastic qualities, it is constantly under tension. This is not a function of the contractile substance but merely of the elasticity of the fibrous tissue and the sarcolemma of the muscle fibers. The passive-stretch curve rises slowly at first, and finally, just before the muscle is torn in two, there is a very steep rise. (Bechtol, C O.: Muscle physiology, Am. Acad. Orthop. Surgeons, Lect. 5:181)

stimulated electrically through its nerve to obtain a maximal tetanus Beginning with the muscle held in a markedly shortened position, no tension is developed As the muscle is stimulated again at increasingly greater lengths, the tension developed by contraction increases rather sharply at first, then after passing a peak lessens somewhat and finally rises sharply again. When the results are plotted on a curve, the curve ascends in an S-shaped fashion. This curve is designated the *total-tension curve* for the muscle.

Next, the experiment is repeated without electrical stimulation of the muscle. The muscle is placed at different lengths, and the various tensions are plotted on the diagram. This curve, in a normal muscle, begins at about the first peak of the total tension curve. and after an initial slow rise it finally rises very steeply in the same region as the second rise of the total-tension curve. Further lengthening of the muscle beyond the point of the steep rise will exceed the limits of elasticity, and the muscle will be torn in two. This is

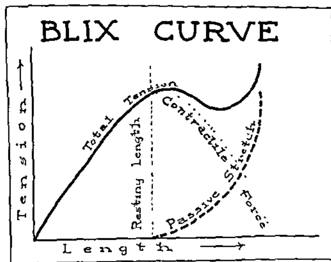


Fig. 22. By subtracting the passive-stretch curve from the total tension curve, it can be seen that the actual contractile force of the muscle is greatest at about the center of its range. This corresponds roughly to the beginning of the passive-stretch curve The peculiar S-shape of the total tension curve is understandable when it is considered as the sum total of the contractile-force curve plus the passive-stretch curve. (Bechtol, C. O.: Muscle physiology, Am. Acad. Orthop. Surgeons, Lect. 5:181)

the muscle and resistance of the muscle to stretch. Myostatic contracture is reversible after short periods of immobilization but becomes increasingly resistant as the immobilization is prolonged. The development of myostatic contracture is dependent upon intact innervation of the muscle. It will not develop if the muscle is deprived of the entire peripheral innervation or the sensory component above (dorsal root). After it is developed it is completely independent of innervation. Myostatic contracture more commonly affects muscles acting over two joints, e.g., the hamstrings, because they are rarely fully stretched by both their joints. The contracture is reversible by prolonged stretching. The passive-stretch curve rises earlier and abruptly because of the fibrous tissue shortening, thereby limiting the range of contractile force. By prolonged stretching, the passive-stretch curve shifts toward the right, and contractile force increases.

When muscle, such as the quadriceps, is bound down by adhesions, it will resist stretching as shown by persistence of the early steep passive-stretch curve. Tendon lengthening by quadricepsplasty will effect a later beginning of the passive stretch.

Immobilization of muscles in a stretched position is said to lead to a slight but permanent loss of power; immobilization in the relaxed position does not result in loss of power.[15] However, it is necessary to study the effect of certain positions upon the shift of the passive-stretch curve before concluding whether or not loss of power is permanent.

Recovery of Function in Partially Denervated Muscles[16]

When one or more spinal roots supplying a muscle are sectioned, the denervation of the muscle is scattered throughout the muscle in a patchy distribution. Recovery in such a muscle is surprising. Denervated fibers in the muscle become reinnervated by remaining intact fibers. This suggests the mechanism by which muscle recovery in poliomyelitis might occur.

The Myoneural Junction

The present concept is of an electrical transmission of impulses across the myoneural junction. It occurs through depolarization of membrane and the various substances such as potassium, acetylcholine, calcium, etc., which transmit the stimulus by altering the process of depolarization. The direct transmission of the impulse by acetylcholine is questionable, since diisopropyl fluorophosphate, an anti-enzyme agent can eliminate all the acetylcholine esterase in the body without materially affecting neuromuscular transmission. The mode of action of curare at the myoneural junction is unknown. It acts to block transmission of impulses. Prostigmine counteracts the action of curare.

Diseases Affecting the Myoneural Junction

Myasthenia Gravis. The basic effect is partial curarization or blocking at the myoneural junction. The condition is improved by prostigmine, while curare, or quinine or procaine makes it worse.

Myotonia Congenita. Unusual sensitivity of the myoneural junction to potassium results in a spontaneous tetanus arising after a voluntary contraction, so that the muscle fails to relax. After several muscular contractions, a more favorable potassium balance is established and after-contracture does not occur. Injection of potassium or prostigmine makes these patients worse. Anything that lowers serum potassium, such as epinephrine, improves the condition.

Family Periodic Paralysis. The muscles are unusually sensitive to a lowered serum potassium level. Injection of potassium effects immediate recovery.

[15] Eisenhauer, J., and Key, J. A. Studies on muscle atrophy, Arch Surg 51 154, 1945

[16] Weiss, P., and Edds, M. R., Jr. Spontaneous recovery following partial denervation, Am J Physiol. 145 587, 1946

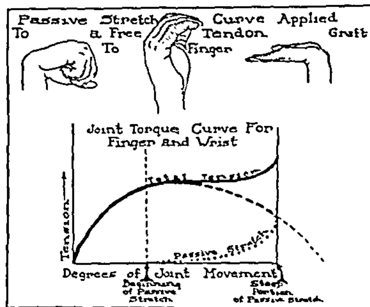


Fig. 24. For the application of a free-tendon graft to a finger, the position of the wrist and the finger joints should be recorded at the beginning of the passive-stretch curve on a normal finger. At the time of surgery, with the wrist and finger in this same position, the beginning of passive stretch of the muscle can be determined directly. The necessary length of the free-tendon graft can then be measured. Surgery and postoperative immobilization can be carried out with the finger and wrist in a greater degree of flexion to prevent tension on the suture lines. (Bechtol, C. O.: Muscle physiology, Am. Acad. Orthop. Surgeons, Lect. 5:181)

ginnings of passive stretch and its steep terminal portion are identified. The beginning of the passive-stretch curve corresponds roughly to the portion of the contractile-force curve where the greatest tension is generated; the steep portion of the passive-stretch curve is the greatest length at which the muscle can act. By correlating these findings with the known range of the joint, the appropriate tendon transplant length can be selected.

The passive stretch and the contractile force for muscles acting about a joint are determined by use of a simple spring dynamometer. The total tension exerted through the range of the joint is called the *joint torque curve*, because it represents the effective torque produced by the muscle through its lever arm. Before a free-tendon graft procedure, the joint torque curve of the finger must first be determined, using one of the patient's normal fingers. Because the profundus tendon crosses both wrist and finger joints, the position of all these joints must be recorded. This can be done simply by sketching the position of the wrist and the fingers at the beginning and the ending of the passive-stretch curve. At surgery, the wrist and the fingers are placed in the position corresponding to the beginning of the passive-stretch curve on the normal finger. The beginning of passive stretch for the muscle to be used is determined by testing its free tendon. Then the necessary length of the free-tendon graft can be measured. The length can be rechecked by placing the wrist and the fingers in full extension and placing the muscle motor under the greatest possible tension, and then remeasuring the length of the graft. Then the actual suturing of the graft can be done with the joints in further flexion to reduce tension upon the suture line. At a later date, after healing, a final determination of the joint-torque curve will indicate if the transplant has been done under the proper degree of tension.

Cineplastic Amputations

In this procedure, an intact muscle supplies the motor power to a prosthesis for the forearm and the hand. It is important that the muscle selected have strong contractile power. The prosthetic length of the cineplastic muscle is the point at which the passive-stretch curve begins. If at the time of amputation the muscle is allowed to retract, its connective tissue will contract over a period of time. As a result, the passive stretch develops earlier and limits the effective contractile range of the muscle. Therefore, if a cineplastic procedure is contemplated at the time of amputation, the muscles to be used as cineplastic motors should be anchored at their normal resting length so that a shift in the passive-stretch curve will not occur.

Myostatic Contracture

If a joint and its muscles are immobilized for a prolonged period of time, limitation of motion of the joint results from shortening of

the muscle and resistance of the muscle to stretch. Myostatic contracture is reversible after short periods of immobilization but becomes increasingly resistant as the immobilization is prolonged. The development of myostatic contracture is dependent upon intact innervation of the muscle. It will not develop if the muscle is deprived of the entire peripheral innervation or the sensory component above (dorsal root). After it is developed it is completely independent of innervation. Myostatic contracture more commonly affects muscles acting over two joints, e.g., the hamstrings, because they are rarely fully stretched by both their joints. The contracture is reversible by prolonged stretching. The passive-stretch curve rises earlier and abruptly because of the fibrous tissue shortening, thereby limiting the range of contractile force. By prolonged stretching, the passive-stretch curve shifts toward the right, and contractile force increases.

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6

Basic Histopathology of Bone

LOCAL FACTORS INFLUENCING OSTEOGENESIS

Compression Forces.[1] Pressure within the physiologic limits of force exerted by the musculature stimulates or enhances osteogenesis. Excessive pressure will cause necrosis with delayed osteogenesis. Absence of compression force fails to stimulate, but does not prevent, osteogenesis. Even in the presence of infection, osteogenesis is stimulated by compression. When stresses are applied to a bone, the trabeculae within that bone develop and align themselves to adapt to these lines of stress (Wolff's law). Pressure force exerted perpendicularly to the axis of a long bone is more likely to cause resorption of bone. Pressure force acting in the line of the bone axis is more likely to cause osteogenesis.

Circulation. A blood supply within certain limits is necessary for osteogenesis. Hyperemia (congestion, sluggish blood flow) results in reduction of osteogenesis. The bone becomes decalcified and osteoporotic. Theoretically, local decreased pH (acidity) effects solution of calcium salts. Deprivation of blood supply results in necrosis of bone. When a section of bone is necrotic, it appears dense in contrast with the surrounding bone which becomes hyperemic and therefore decalcified.

Disuse, Inactivity. Osteogenesis is reduced, and all bone in the immobilized part becomes osteoporotic. Sluggish blood flow and lack of muscle forces are the main causative factors.

Inflammation. Injury or infection causes a reactive hyperemia which in turn effects dissolution of calcium salts. Leukocytic proteolytic ferments interfere with formation of osteoid.

[1] Eggers, G. W. N., Shindler, T. O., and Pomerat, C. M.: Osteogenesis. Influence of the contact-compression factor on osteogenesis in surgical fractures, J. Bone & Joint Surg. 31:693, 1949.

Neurotrophic Disturbance (Sudeck's Atrophy; Reflex Sympathetic Dystrophy). Following trauma, a reflex vasodilatation causes widespread osteoporosis which typically is spotty in appearance.

Traction Stress. This force tends to reduce bone formation. For example, distraction at a fracture site interferes with union.

Shearing Stress. When two bony surfaces are subjected to shearing stresses, osteogenesis is held to a minimum, and fibrous tissue and cartilage formation are encouraged.

The Periosteum. The presence of the enveloping periosteum is necessary for osteogenesis. For example, when a rib with its periosteum is removed, it does not regenerate. Periosteum, when transplanted, will continue to grow bone. Destruction of the periosteum, as by thermal cauterization, retards osteogenesis. When the periosteum is stripped upward, as by a tumor, a layer of new periosteal bone forms beneath it.

Availability of Local Bone Substance. In certain situations osteogenesis requires that bone materials be available in the immediate neighborhood. Calcium salts alone are insufficient as demonstrated by the fact that injection of mineral salts into the site has no effect on bone formation. In the healing of a fracture, the adjacent living bone is resorbed and redeposited in the callus. When a fibrous union exists between fragments, a bone transplant laid alongside the pseudarthrosis will effect ossification within the fibrous tissue. Therefore, it is unnecessary to remove the fibrous tissue in order to effect a bony union.

REPAIR OF FRACTURES

Interruption in continuity of bone, whether traumatic or surgical, is followed by a definite histologic sequence aimed at bridging the defect. A basic understanding of this process is

from the opposite fragment. Between the fragment ends, where constant shearing motion is inevitably present and circulation is deficient, considerable cartilage forms. This tissue extends outward from between the fragments and becomes continuous with the fibrocartilage of the external callus. Like the latter, it too must be replaced by metaplasia and endochondral ossification.

The small amount of necrotic bone over the fragment ends is replaced by the process of creeping substitution. The dense living cortical bone about the fracture site is resorbed and remodeled by osteoclasts and replaced by spongy bone.

The shaft of a long bone generally unites mainly by formation of external callus. The new bone expands and extends as an arch which bridges the fracture gap. From this fixed arch bridge, new trabeculae extend centripetally between the fragment ends and join endosteal trabeculae. By the process of remodeling, primary cancellous trabeculae are absorbed, and new lamellae are laid down along lines corresponding to direction of stress, usually parallel with the long axis of long bones. Haversian systems are constructed by lamellar plates laid down concentrically about blood vessels. Cortical walls are restored, the medullary canal is re-established, and fatty or hematogenous cellular elements fill the marrow spaces.

The blood supply of callus is derived from new subperiosteal and metaphyseal perforating arteries and the nutrient artery. Blood calcium, phosphorus and alkaline phosphatase are not altered significantly during fracture healing.

DELAYED UNION AND NONUNION[6]

Bone defects as a result of trauma or surgically induced may heal slowly or fail entirely to heal. *Delayed union* is an arbitrary term implying that union is taking place over a

[6] Urist, M. R., Mazet, R., and McLean, F. C.: The pathogenesis and treatment of delayed union and nonunion, J. Bone & Joint Surg. 36-A 931, 1954.



FIG. 25. Fracture callus. A cancellous type of bone forms adjacent to the cortex, replaces the cartilage and extends distally as a bridge to meet the callus formation from the opposite fragment. The other fragment is not seen in this section.

longer period than is customary. *Nonunion* defines complete failure of healing of a fracture. It is characterized by (1) a bone defect, (2) false motion, (3) sclerosis of the bone ends, (4) rounding, mushrooming and molding of the fracture surfaces, and (5) sealing of the medullary canal with compact bone.

Causes of Delayed Union and Nonunion

The principal causes are:

1. Gaps caused by displacement, comminution, loss of bone substance, and soft tissue interposition.
2. Extensive surgical dissection causing loss of vital soft tissue attachments. Such freed fragments become necrotic and act like free bone grafts which must be slowly replaced by new bone.
3. Infection.
4. Metallic internal fixation devices which cause bone necrosis.
5. Inadequate circulation, e.g., in the neck of the femur.
6. Metabolic disturbances.
7. Inadequate immobilization.

Certain bones characteristically form very little callus, and union is normally very slow. The lower third of the tibia is an example. Therefore, it is easily disturbed by the factors which interfere with union. On the other hand, other bones display a strong tendency toward union in spite of adverse conditions, e.g., the ribs.

Normal Healing of a Fracture[7, 8]

Knowledge of histophysiology of fracture healing is basic to an understanding of the events leading to nonunion. The repair process begins by proliferation of periosteum and the endosteum at some distance from the line of fracture, the newly formed tissue extending into the fracture site. Within the periosteum intramembranous ossification takes place, producing cancellous bone. Endosteal new bone is appositional. Within the proliferated periosteum and between the fracture fragments, fibrocartilage is differentiated. Its appearance is a necessary preliminary to bridging of the fracture by bone. The fact that perivascular young connective tissue cells rapidly invade and replace the fibrocartilage suggests that the latter possesses some physicochemical effect which induces osteogenesis. Transplants of fibrocartilaginous tissue will similarly induce invasion of osteogenetic connective tissue cells from host tissue. Therefore, the healing of a fracture takes place by (1) proliferation of osteogenetic cells of the periosteum and the endosteum, and (2) by ingrowth of perivascular young connective tissue cells which differentiate into osteoblasts under the influence of fibrocartilage.

The Structure of Callus in Ununited Fractures

Within the first few months, the tissue resembles normal callus. The spindle-shaped capsule of proliferated periosteum envelops the fracture site, and cancellous new bone forms on the periosteal surface of the bone at a distance. Endosteal new bone forms behind the fracture surfaces. Fibrocartilage covers the bone ends. However, in ununited fractures, an amorphous material consisting of fibrinoid, hyaline and mucinous fluid accumulates within the fracture interval. This material is surrounded by patches of necrotic connective tissue and round cells.

Fibrinoid is the principal constituent of the amorphous material. It is acellular, homogenous, refractile and stains metachromatically with toluidine blue. It can be reproduced experimentally by mechanical trauma which causes extravasation of plasma into normal connective tissue.[9] The mechanism of its formation is believed to be precipitation of mucopolysaccharides from the ground substance by substances derived from necrosis of tissue. The study of this substance is important to understanding the pathogenesis of connective tissue [illegible]

[illegible] observed. The accumulation of mucinous fluid is pronounced when excessive motion between

[7] Urist, M R, and Johnson, R. W Calcification and ossification IV Healing of fractures in man under clinical conditions, J Bone & Joint Surg. 25: 375, 1943

[8] Urist, M. R, and McLean, F. C.· The local physiology of bone repair, with particular reference to the process of new bone formation by induction, Am. J Surg. 85 444, 1953.

[9] Wu, T. T.: Über Fibrinoidbildung der Haut nach unspezifischer Gewebsschadigung bei der Ratte, Virchows Arch. path Anat. 300:373, 1937.

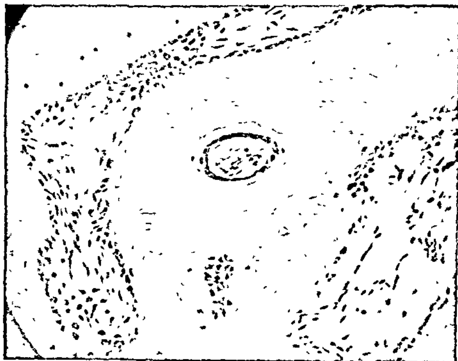


PLATE 2. Aseptic necrosis and creeping substitution. Active osteogenesis and osteoclastic resorption are taking place at the same time. Loose vascular osteogenetic granulation tissue occupies the marrow spaces and the enlarged haversian canals. A concentric lamella is being laid down in the haversian canal at the center of the section. (× 160)

fragments is present The viscous fluid stains metachromatically with toluidine blue. One may postulate that the trauma increases capillary permeability, and the extravasated fluid combines with the fibrinoid material to form mucinous fluid.

Patches of another eosinophilic substance, hyaline, are seen It lacks the granular or fibrillar appearance of fibrinoid and does not stain metachromatically with toluidine blue Like fibrinoid, it increases in quantity with the age of the ununited fracture.

Elsewhere, dense scar tissue occupies the fracture interval. It is increased in amount in the presence of infection.

With the passage of time, the medullary cavities at the bone ends become obliterated by dense cancellous and compact bone. One bone end, usually the proximal, becomes expanded and develops a concave surface which articulates with the convex surface of the opposite bone end. The surfaces are covered with

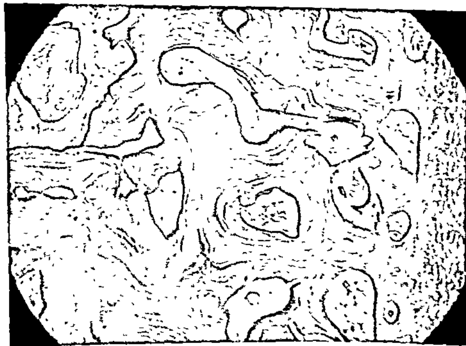


PLATE 3. Radiation necrosis of cancellous bone. The characteristic "molasses" appearance is well shown. Marrow spaces are empty or contain fatty tissue. (× 35)



Plate 4. Radiation necrosis of cortical bone. This displays the characteristic osteolytic defect, which is filled with loose, relatively avascular fibrous tissue. The margins of the defect are sharp and devoid of cells. Haversian canals contain a peculiar dark-staining substance. (× 25)

hyaline and fibrocartilage. They are joined by fibrous connective tissue containing amorphous fibrinoid and hyaline material and a false joint cavity containing a viscous fluid. A continuous increase in the quantity of new bone is laid down from the rim of the concave bone end.

Mechanism of Production of a Nonunion

The connective tissue is injured and rendered necrotic as a result of inflammation, infection, motion and friction. This results in degeneration of ground substance with production of fibrinoid and mucinous fluid containing polysaccharides. The process is halted and reversed by immobilization, the defect becoming filled with new fibrocartilaginous tissue and new bone.

Treatment

Before embarking upon a course of treatment, one must first determine whether actual nonunion or a state of delayed union exists. It is possible that additional immobilization is all that is necessary. Watson-Jones states that nonunion is rare, and continued immobilization will effect a union in almost all cases. However, an extended period of incapacity may be economically disastrous, and an effort to expedite recovery should be considered in every case of delayed union.

The attack is directed at eliminating the causative factor. First, infection should be eradicated. The main fragments should be approximated closely and some compression effected to promote osteogenesis. A wide gap between fragments requires bridging by a cortical graft. Mobility between fragments is overcome by fixation by a metallic appliance (e.g., an intramedullary nail) or by affixing a cortical graft to both fragments. The cortical graft and multiple cancellous bone chips about the site of pseudarthrosis induces bony bridging. It is unnecessary to excise the soft tissue from the pseudarthrosis interval. Osteotomizing the associated bone, such as the fibula in the case of an ununited tibia, will permit closer approximation of the fragments. In certain situations, externally applied bone grafts are not feasible. An ununited navicular bone of the wrist requires an intramedullary bone peg which provides fixation. Finally, plaster cast immobilization is continued until roentgenographic examination demonstrates bony bridging of the interval.

ASEPTIC NECROSIS OF BONE

Death of bone as a result of deprivation of circulation is designated as *aseptic necrosis.*

Causes

Any condition which shuts off the blood supply will produce aseptic necrosis. Causes

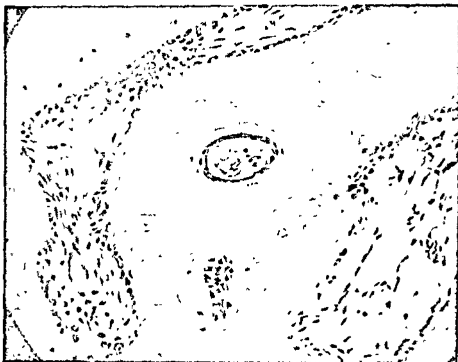


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include fracture, dislocation, extensive stripping of periosteum from bone during surgery, caisson disease and exposure to the roentgen ray and radioactive substances. By definition, infection does not cause aseptic necrosis; but, secondarily, accumulation of exudate within unyielding bony walls shuts off the blood supply and contributes to necrosis in osteomyelitis.

Sites of Predilection

The most common locations are as follows:

Femoral Head. The blood supply enters by way of capsular vessels which enter the femoral neck distally and pass centrally to the head. A fracture through the femoral neck, especially in the proximal portion of the neck, or a severe dislocation which ruptures the capsular vessels is likely to produce aseptic necrosis of the femoral head.

Carpal Navicular. The blood supply enters the bone mainly at its distal portion. The more proximal the fracture, the more likely is circulation to be interrupted to the proximal fragment.

Astragalus. The main nutrient vessels enter the bone at its neck. Extensive stripping about this site or a complete dislocation is often followed by aseptic necrosis.

Segmental Fractures. A large or a small fragment is often separated from its source of nutrition and undergoes necrosis.

Other Locations. The capitellum of the humerus, the radial head and the lateral femoral condyle at the knee are less common sites.

Microscopic Appearance[10, 11]

The bony architecture is unaltered, trabeculae retaining their original contour. However, the lacunae are empty, and the marrow contains formless debris. No evidence of vascularity or cellular activity can be found.

The adjacent bone which has retained its blood supply undergoes reactive hyperemia in preparation for replacement of the dead tissue. Vascular congestion effects resorption an thinning of osseous structure which is likewis encouraged by inactivity. This produces th characteristic demineralization and trabecula thinning in bone which lies adjacent to ne crotic bone. The latter, lacking in blood supply cannot undergo bone atrophy and therefor retains its original density and architecture By contrast with hyperemic bone, it appear to be very dense, but actually its density i unchanged.

Capillaries with a young fibrous stroma penetrate the marrow spaces and enter the haversian canals within the necrotic tissue. Phagocytes remove debris. Osteoclasts resorb dead bone, thinning the dead trabeculae and widening the haversian canals. At the same time that bony resorption is taking place, osteoblasts lay down osteoid seams, trabeculae being entirely replaced by new bone (creeping substitution). After each haversian canal has been widened sufficiently, new layers of lamellar bone are laid down within the canal, and the haversian system is reconstituted.

Roentgenographic Characteristics

The uninvaded dead bone lacking blood supply cannot alter its structure and retains its original density. The adjacent living bone [illegible]

The dead bone appears to be relatively dense, although it actually is unchanged.

A fracture which deprives a segment of bone of its blood supply will unite slowly even though one fragment is necrotic. At the line of juncture between necrotic and living bone is observed osteoid and new bone. This forms a point of weakness through which fracture may occur. Therefore, whenever union develops between living and dead bone, protective immobilization should be enforced until complete replacement is demonstrable by roentgenogram.

Articular cartilage overlying necrotic bone does not itself become necrotic because it derives its nutrition from synovial fluid. However, when subchondral fractures occur as necrotic trabeculae yield to weight-bearing pressures, abnormal mechanics lead to degenerative arthritis.

[10] Phemister, D. B.. Lesions of bones and joints arising from interruption of circulation, J Mt Sinai Hospital 15:55, 1948.

[11] ———: Repair of bone in the presence of aseptic necrosis resulting from fractures, transplantations, vascular obstruction, J. Bone & Joint Surg. 12:769, 1930.

riod of months, from 90 to 99.9 per cent of radium is eliminated. A minute amount of radium is deposited intraskeletally and is eliminated less easily. This residual radium can be measured by a scintillation counter.

Location of Radium in Bone

The presence and the position of radioactive substance in tissue can be determined by autoradiography. A sensitized photographic film applied to the tissue surface will exhibit markings characteristic of radiations.

Radium is found as small highly concentrated focal spots in both cortical and cancellous bone Heavy and uniform concentrations are observed at the epiphyses, especially in the subchondral area.

Histopathology

The late effects of irradiation are characterized by:

1. **Necrosis of Bone With Atypical Architecture.** This is usually observed at the epiphyses, especially below and adjacent to articular surfaces. The trabecular bone displays peculiar undulating contours, an appearance likened to "molasses candy." Lacunae are empty. Marrow spaces are filled with loose acellular fibrous tissue. Osteoblastic activity is lacking. This necrotic bone exhibits no evidence of radioactivity. Compact bone likewise is necrotic as evidenced by absence of osteocytes The haversian canals are widened and often filled with an amorphous material staining dark blue by H-E stain.

2. **Areas of Destruction in Compact Bone.** In cortical bone, sharply circumscribed areas of bone destruction are replaced by loose relatively acellular fibrous tissue. The margins of bone enclosing the osteolytic area appear to be eaten away, but no osteoclasts are present. These osteolytic cavities produce roentgenographic areas of decreased density which appear as streaks in long bones and as punched-out spots in the skull and the long bones. Often the dark-staining amorphous material occupies these defects as well as the enlarged haversian canals.

An area of bone which has undergone radiation necrosis is either sequestrated and slowly absorbed, or is slowly replaced by creeping substitution.[15] Regeneration of bone is dependent upon the state of the overlying tissues. Periosteal blood vessels are often destroyed.

Clinical Picture[16]

The acute radiation syndrome is characterized by progressive weakness, infection, necrosis, sloughing and bleeding of the mouth and the pharynx, large purpuric areas, prostration, delirium, and death from infection, pancytopenia and collapse.

The chronic radiation syndrome is more important to the orthopaedic surgeon. Symptoms appear after 15 years, arise from destructive changes in the skeleton, are not severe and are *not progressive. Skeletal changes* usually develop in weight-bearing bones or those subjected to repeated trauma. Vertebral bodies undergo spontaneous collapse. Pathologic fracture of a long bone almost always occurs in the femoral shaft. Although pain is the predominant complaint and may be localized to any skeletal site, pathologic fracture often takes place without antecedent symptoms. The fracture unites slowly. Aseptic necrosis of the femoral head presents the typical picture of degenerative arthritis of the hip. Involvement of maxilla and mandible is very frequent but not as severe as in acute cases. It develops regardless of the mode of intake of the radioactive substance. A very characteristic dental change is a reduction of density which gives a pinkish hue to the teeth. Death occurs from severe anemia, crippling bone lesions and malignancy.

Roentgenologic Findings

These consist of the following:

1. **Areas of Decreased Density.** Small osteolytic defects measuring 1 to 2 mm. by 5 20 mm. produce streaks in long bones punched-out lesions in the skull and the bones.

2. **Areas of Increased Density.** T' produced by atypical necrotic bo found in relation to the areas of

[15] Phemister, D. B.: Radiation nec Am. J. Roentgenol. 16:340, 1926.

[16] Martland, H. S.: The occurrence in radioactive persons, Am. J. Cancer

[illegible] Mottled moth-eaten densities appear [illegible]often in the femoral head, the humeral head, the glenoid process and the bones of the foot. When it is found concentrated about [illegible]r and inferior borders of vertebral bod[illegible] collapse of the vertebrae is common. No periosteal reaction is observed. The skeletal changes are slowly reversible with passage of [illegible]erable time.

Diagnosis

[illegible]septic necrosis of the femoral head, the [illegible]eral head, the glenoid process and the [illegible]es of the foot, and pathologic fracture of [illegible] vertebral body or a femoral shaft should [illegible]ggest the possibility of irradiation necrosis. [illegible]e diagnosis may be determined by:

1. History of radium inhalation, ingestion, [illegible]ministration.
2. X-ray changes.
3. Measure radon in expired air. Radon, the first daughter element in the decay of radium, is mainly eliminated in the breath.
4. Radiochemical analysis of excreta (24-to-48-hour specimen).
5. External measurement of gamma-ray activity.
6. Radiochemical analysis or autoradiography of biopsy specimen. Viable bone is more likely to exhibit radioactivity than necrotic bone.

Postradiation Sarcoma of Bone[17]

A latent period averaging about 10 years exists between the time of exposure to radiation and appearance of the neoplasm. Sarcoma may develop in a normal bone which lies in a field of soft-tissue irradiation. More commonly, radiation has previously been applied to a benign type of bone lesion. The onset of neoplastic activity is acute, and development of the tumor is rapid. A bone sarcoma may develop regardless of whether single or multiple exposures have been employed. Giant cell tumors are particularly prone to malignant degeneration. These facts seem to suggest that benign bone lesions must never be irradiated unless absolute evidence of malignancy is obtained, and then should be used only as an adjunct to amputation.

[17] Sabanas, A. O, Dahlin, D. C., Childs, D. S, and Ivins, J C.: Postradiation sarcoma of bone, Cancer 9 523, 1956.

CAISSON DISEASE
(Decompression Sickness; "The Bends"; Aero-embolism)

Atmospheric nitrogen is absorbed through the lungs and deposited by the blood in various body tissues, particularly those containing fat (bone marrow) and lipoid substances (brain, spinal cord). At greater than atmospheric pressure, nitrogen absorption is proportionately greater. When atmospheric pressure is suddenly lowered in rapid ascent to a higher altitude, as experienced by divers, tunnel workers, or pilots of nonpressurized aircraft, nitrogen gas is liberated in a concentration which cannot be absorbed readily by the blood stream or excreted quickly by the lungs. As a result, gas bubbles accumulate in the tissues, causing direct pressure asphyxiation, and in the blood stream, producing vascular occlusion and infarcts.

Symptoms, which develop within minutes to a few hours, depend upon the parts of the body affected. Complaints include severe pain about the joints of the extremities ("the bends"), vertigo ("the staggers"), and various neurologic manifestations. Prompt treatment will often effect resolution of symptoms. Delay in treatment allows tissue necrosis to develop and permanent sequelae are produced, especially paralyses and extensive skeletal lesions. In severe untreated cases, circulatory failure, coma and death quickly ensue.

Skeletal Lesions[18-20]

Bones of the extremities which are rich in fatty marrow may undergo extensive necrosis. The large lesions are usually found in the diaphysis and may extend to the epiphyses. Infrequently, they are limited to the epiphyses. They are symmetrical and are found most often in the lower extremities. The de-

[18] Coley, B. L, and Moor, M.: Caisson disease in bones and joints, Am. Surg. 111:1065, 1940.

[19] Kahlstrom, S. C., Phemister, D. B, and Burton, C. C.: Bone and joint changes in caisson disease, Surg., Gynec. & Obst. 68:129, 1939.

[20] Walker, W. A. Aseptic necrosis of bone occurring in caisson disease, J. Bone & Joint Surg. 22.1080, 1940.

gree of involvement varies from small localized areas, which are often generally distributed, to vast individual areas of necrosis of long bones Small areas of aseptic necrosis may exist throughout the skeleton without being evidenced on gross inspection or by roentgenogram. These are gradually replaced by new bone. A large area of necrosis usually involves the medulla and may include an extensive portion of the diaphyseal cortex. Gradually, over a considerable period of time, by the process of creeping substitution, the periphery of dead bone and marrow is resorbed and replaced However, after a variable period of time, the process of replacement slows down or ceases entirely, and the dead area no longer decreases in size. Instead, calcium deposits are laid down in the interior, increasing the density. A narrow zone of dense ossification at the periphery sharply delimits the lesion. Thus at the late stage, years after the onset, the characteristic roentgenographic shadow of a persisting lesion is a large blotchy area of increased density with a narrow undulating margin of particularly heavy density.[21]

Death of trabeculae is more widespread than is evident on roentgenograms because the majority of infarcts are of microscopic size. Eventually and characteristically, bone fragility becomes manifest at large joints where subchondral bone succumbs to pressure, and severe degenerative arthritis is produced. Following exposure, after an asymptomatic interval of several years, the symptoms of severe deforming arthritis first appear, typically affecting one or more joints of hips, knees and shoulders, and pertinently excluding the spine. Vertebrae contain large amounts of vascular cancellous bone and therefore are unaffected by caisson disease.

Pathologic fracture has not been reported.

TRANSPLANTATION OF BONE

Removing bone from one site and transplanting it to another is a common surgical procedure. It is employed (1) to replace bone substance lost through injury or disease and (2) to bring about bony union.

[21] Hodges, P. C., Phemister, D. B., and Brunschwig, A.: The Roentgen Ray Diagnosis of Diseases of Bones and Joints, p. 222, New York, Nelson, 1938.

The use of the term "bone graft" is controversial, since microscopic examination of such bone reveals empty lacunae, suggesting death of osteocytes and matrix. Further, transplanted bone is invaded by blood vessels from surrounding living bone which enter the marrow and the haversian canals, is resorbed on its surfaces and from within the canals and is replaced by trabeculae of new bone. The transferred bone acts as a bridge across which new bone may form. By its presence it stimulates new bone formation, perhaps by providing a supply of necessary minerals and proteins. This is readily demonstrated in the case of fibrous union of bone fragments. By placing a segment of bone adjacent to the site of nonunion, bony trabeculae are encouraged to bridge the bony defect.

Therefore, the bone transplant serves several functions.

Immobilization. For this purpose, dense cortical bone is used. Most often it is removed from the subcutaneous aspect of the tibia and is fixed in place by metallic screws. Cortical bone is resorbed and replaced very slowly, and prolonged protection from weight-bearing is necessary.

Osteogenesis. Replacement by new bone is active and rapid when cancellous bone is used. Its soft marrow is quickly penetrated by capillaries, and many of the original osteoblasts survive and continue to produce new bone The extensive network of fine trabeculae provides a tremendous surface area about which new bone is laid down while the slender spicules of necrotic bone are rapidly absorbed. The iliac bone is an excellent source of cancellous bone.

Replacement. Extensive loss of the shaft of a long bone occasionally may be replaced. Most commonly, the upper half or two thirds of the fibula may be utilized.

Microscopic Findings

When bone is removed from its site of origin, the osteocytes disappear, and empty cell spaces suggest death of the bony matrix. When such bone is immediately transplanted to a bed of adequate nutrition, particularly when the bone is cancellous, the soft tissue elements between the trabeculae survive and provide osteoblasts which envelop the trabeculae and

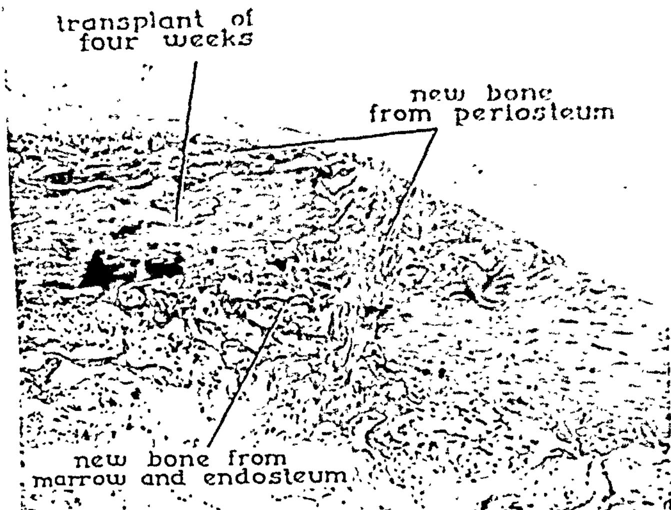


FIG. 26. Transplanted bone, four weeks old. The transplanted bone is necrotic as shown by empty lacunae. It originally was a solid cortical graft which is now porous by virtue of resorption at its surface and from within the haversian canals which now appear widened. New trabeculae of bone formed from the periosteum and endosteum of the host bone have spread about and become firmly fixed to the necrotic bone. Where new living bone lies in contact with necrotic bone, the latter is being resorbed by the process of creeping substitution. (Ham, A. W.: Histology, ed. 2, Philadelphia, Lippincott)

surround them with newly formed bone while the necrotic bone is resorbed (by creeping substitution). This process is facilitated by medullary elements and capillaries entering the implanted bone from the surrounding host bone. The spongiosa contains a superabundance of these elements which can invade and supply osteoblasts. Therefore, transplantation of bone is more effective in producing new bone when placed in a bed of cancellous bone. When the transplanted bone consists chiefly of dense cortex, marrow spaces are lacking, and haversian canals are inadequate for penetration by osteogenetic granulation tissue. The thick bone must be entirely enveloped and slowly absorbed and replaced at its periphery. Only a small amount of resorption takes place by vessels which penetrate the haversian canals. Osteogenesis and replacement are slow. Therefore, cancellous bone seems best suited for transplantation when osteogenesis is the major consideration. It is readily invaded, and its trabeculae are rapidly resorbed and replaced. Rapid revascularization occurs, and many of the original osteoblasts survive. Cancellous bone containing red marrow is revascularized better than that containing fatty marrow.

When a homograft is implanted, its medullary elements quickly disappear, probably because of protein incompatibility with the host tissue. Therefore, the supply of osteoblasts

must originate from the invading tissue. Microscopically, this is demonstrated by the fact that, although soft, cancellous, easily penetrated bone is used, new bone formation and creeping substitution are more active at the periphery of the implanted bone and declines in degree as the center is approached. Therefore, homogenous bone is incorporated more slowly than autogenous bone, the difference depending on the degree of protein incompatibility. Heterogenous bone, when examined microscopically, is entirely incompatible with human bone. It becomes enveloped with a capsule of connective tissue and is gradually resorbed. Replacement with new bone also occurs very gradually from the periphery but not by creeping substitution. These facts indicate that autogenous bone is best for transplanting, and spongiosa constitutes the most favorable bed.

The Bone Bank

Recently, the use of homogenous bone has greatly increased by improvements in methods of preservation. The desirability of having large amounts of bone available at operation is obvious and tempting. Bone removed in the course of surgical procedures on clean cases and amputation specimens is preserved by freezing or storing in antiseptic solutions. Reports to date seem to be very favorable regarding the results of this method

Certain facts argue against the use of preserved bone. Homografts are not readily invaded, even when placed in a bed of adequate nutrition. Occasionally, such bone excites a foreign body response with outpouring of exudate and a febrile reaction, which does not subside unless the bone is removed. This suggests protein incompatibility with the host tissue. There is no reason to expect bone, a protein material, to react differently than blood or skin when transferred from other individuals. The possibility of contamination and transfer of disease, with its medicolegal implications, should be borne in mind.

Autogenous bone should be used whenever possible. The added risk of obtaining the bone is negligible, and sources of supply are plentiful. The only justification for the use of preserved bone is in the very young and the aged and where the demands of the surgical procedure require large quantities of bone.

Isotope Studies of Bone Transplants[22]

Revascularization of bone transplants may be determined by implanting them in the experimental animal and injecting the latter with radioactive phosphorus. The ability of a bone graft to take up P^{32} is determined by the count/minute/gram of tissue. The highest counts are observed in fresh autogenous transplants and the lowest in stored homologous bone grafts. Of course, the take-up of P^{32} by a homologous transplant is related to developing blood supply which, in turn, depends on tissue compatibility between graft and host.

[22] Kiehn, C L, and Glover, D. M · Study of revascularization of stored homologous grafts by means of radioactive phosphorus, Plast. & Reconstruct Surg 12,233, 1953.

PART TWO

General Orthopaedic Conditions

7

Metabolic Bone Disease and Related Dysfunction of the Parathyroid Glands

FACTORS AFFECTING CALCIUM METABOLISM

The deposition of calcium is favored by:

1. Availability in food
2. Alkalinity of serum
3. Vitamin D

Resorption from bone is favored by:

1. Acidosis
2. Hyperparathyroidism
3. Hyperthyroidism
4. Pregnancy or lactation
5. Starvation—acidosis, lack of Ca, P, vitamin D
6. Excessive vitamin D
7. Chemical poisoning—strontium, magnesium
8. Excessive diuresis
9. Local causes—inflammation, vascular congestion
10. Inactivity, immobilization—leads to excessive removal of calcium, hypercalcuria, renal stones
11. Injury (Reflex vasomotor mechanism? Selye adaptation syndrome?)

Absorption from intestine favored by:

1. Acidity of digestate. In the distal intestine, alkalinity precipitates the calcium
2. Vitamin D
3. Bile salts (to split fats completely and prevent soap formation)

Absorption from the intestine is hindered by:

1. Alkalinity—alkalies, achlorhydria
2. Excessive phosphates
3. Excessive fatty acids—form insoluble calcium soaps
4. Excessive carbonates—form insoluble calcium carbonate

Availability from blood stream depends on its ionization:

1. Colloidal calcium is bound to protein, therefore unavailable
2. Salt of $Ca_3(PO_4)_2$ in solution exists in equilibrium with
3. Ionized calcium

Solution of calcium in serum is favored by:

1. Decreased pH
2. Protein concentration
3. Lowered serum ionic strength
4. Magnesium and strontium
5. Increased parathyroid activity

PARATHYROID GLANDS

Anatomy

The parathyroid glands are very small structures about 10 × 5 mm. in size, flattened and ovoid in shape, yellow brown in color, approximately 4 in number, and lying in relation to the posterior aspect of the thyroid gland. Occasionally, the inferior pair may be situated in the mediastinum. As many as 12 glands may be present.

Histology

Up to the age of 10, the gland is composed of a uniform type of cells densely packed as a continuous mass or anastomosing cords; less commonly, they are arranged as follicles with a colloid material at the center. These cells are designated *principal or chief cells.* They have a large, vesicular, centrally placed nucleus and a faintly staining homogeneous cytoplasm. At about the age of puberty *oxyphil cells* appear. These are larger than the principal cells, have a smaller, darker-staining nucleus and a deeply acid-staining cytoplasm.



Fig 27. (*Left*) Normal parathyroid gland. Note the adipose tissue and vascularity throughout the gland. (*Right*) Adenoma of the parathyroid gland. Note the encapsulation separating the adenoma from the normal gland. Because adenoma causes hypercalcemia, nephrocalcinosis, renal insufficiency and retention of serum phosphorus, parathyroid hyperplasia takes place in response to hyperphosphatemia. This hyperplasia takes place in all the parathyroid tissue, including that which surrounds the adenoma, as seen in this section.

Table 1. Normal Values Serum Calcium and Serum Phosphorus

	Infants	Children	Adults
Calcium (mg per 100 cc.)	10 5–12 0	10 0–11 5	9 5–10 5
Phosphorus (mg. per 100 cc)	5 5–6 5	4.5–5 5	2 5–4 0

Table 2. Normal Values for Serum Phosphatase*
Units per 100 cc of Serum

	Acid (pH 5 0) Adults	Alkaline (pH 8 6–9 3) Adults	Alkaline (pH 8 6–9 3) Children
Bodansky†	0 0–0.4	1 5–4 0	5 0–12 0
King-Armstrong‡		3 7–13 1	15 0–20 0
King-Armstrong‡ (Gutman's Modification)	0 3–3 25		

* Sunderman, F W Am J. Clin Path 12 404 (1942)

† Substrate—sodium B-glycerophosphate (unit is based on 1 mg P)

‡ Substrate—di-sodium phenylphosphate (unit is based on 1 mg Phenol).

Physiology

The parathyroid secretes a hormone with the following functions:

1 *It maintains the serum calcium level.* When the serum calcium is low, as from dietary lack, the normal level is restored by dissolution and osteoclasis of bone. The low serum calcium causes parathyroid hyperplasia. Conversely, when serum calcium is high, parathyroid hypoplasia results; the excess is excreted and deposited in bone.

2. *It lowers the serum phosphorus level.* Parathyroid hormone encourages excretion of phosphorus by inhibiting renal tubular resorption. When the serum phosphorus is high, as in low calcium, high phosphorus rickets, parathyroid hyperplasia develops in an effort *to eliminate the excess.*

3. *It increases diuresis of both calcium and phosphorus,* mainly by inhibiting tubular resorption.

4. *It stimulates osteoclastic resorption of bone.*

5 *The hormone will directly effect dissolution of bone.* When a gland is placed in direct

contact with bone, the latter will absorb although osteoclasts are not yet apparent

6. It inhibits the calcifying effect of vitamin D.

7. It increases the solubility of calcium and phosphorus, maintaining these substances in ionic form beyond their expected solubilities

In studying the following metabolic diseases, it is important to determine the part played by the parathyroids Increased parathyroid secretion can be primary or secondary Secondary hyperplasia develops as a response to serum calcium and phosphorus levels An elevation of blood phosphorus increases parathyroid secretion, as does also a relative lowering of serum calcium The parathyroid hormone will then effect osteoclast formation and osteoclasis and also direct dissolution of bone When clinical evidence of parathyroid hyperfunction exists, a low serum calcium or high phosphorus signifies a secondary hyperplasia of the glands, and the cause must be sought, e.g., diet, renal insufficiency, etc When serum calcium is high and phosphorus low, primary parathyroid adenoma or hyperplasia is the cause.

Kidney Function: Relation to Secondary Hyperparathyroidism

Normal kidneys can eliminate phosphorus easily When nonfunctioning glomeruli form a barrier to passage of phosphorus, the hyperphosphatemia urges the parathyroids to greater activity in an effort to excrete the mineral, resulting in excessive excretion of parathormone. A hyperparathyroid state is created. The excess parathormone increases the rate of bone absorption with consequent rise of calcium and phosphorus in the blood stream. Since these elements cannot be eliminated by the kidneys, they are deposited throughout the soft tissues. Deposition in the kidneys themselves forms multiple staghorn calculi Metastatic pathologic calcification occurs everywhere, including the walls of blood vessels. The large bowel excretes the excess of calcium and phosphorus, although inadequately. All types of kidney diseases and congenital anomalies basically act the same way. Renal rickets eventually results in the child; renal osteomalacia, in the adult In children the changes are influenced by the presence of actively growing epiphyseal cartilage plates. These plates and the osteoid tissue just beyond becomes excessively wide and irregular. This is a high phosphorus rickets, in contrast with the usual normal or low phosphorus rickets caused by deficiency of vitamin D.

The main features are:

1 Marked renal insufficiency of long duration
2 Phosphate retention with high serum phosphorus
3. Slight reduction of serum calcium.
4 Marked acidosis
5. Metastatic calcium deposits near joints
6 Monckeberg sclerosis
7. Osteitis fibrosa generalisata
8 Enlargement of parathyroid tissue

Treatment consists of lowering the phosphorus intake, reducing its absorption by aluminum hydroxide, and efforts directed against the kidney disease. When kidney disease is primarily tubular, loss of calcium is excessive and large quantities of vitamin D and calcium must be administered.

PRIMARY HYPERPARATHYROIDISM

(Osteitis Fibrosa Cystica; von Recklinghausen's Disease; Parathyrotoxicosis)

When secretion of parathormone becomes excessive, it is reflected in bone by a marked increase of osteoclasts, rapid resorption of bone, decrease of osteoblasts, and fibrous replacement of marrow. Both calcium and phosphorus are thrown into the blood stream; although both are excreted mainly by the *kidneys*, elimination of phosphorus is accomplished more readily with the result that the blood calcium is elevated and phosphorus is lowered. Alkaline phosphatase is elevated supposedly because of a compensatory effort at restoring the resorbed bone.

Pathology.[1] Most frequently, an adenoma measuring up to 6 cm. in diameter is situated in one of the parathyroid glands. Less often 2 adenomata or diffuse hyperplasia is the cause of excess secretion. The adenoma is composed mainly of the pale, clear, chief or principle cells. Rarely, oxyphils form the *main compo-*

[1] Luck, J V Bone and Joint Diseases, Springfield, Thomas, 1950.

nents. The cells tend to form acini, cords and patternless masses. Normal gland tissue and hyperplasia are characterized by cell uniformity.

Skeletal changes are generalized and include the following:

1. *Diffuse Bone Resorption.* Large numbers of multinucleated osteoclasts are observed in Howship's lacunae, Haversian canals are enlarged, and cortices are transformed to cancellous bone of paper-thin thickness.

2. *Deformities.* Long bones bend under the stress of weight-bearing. Intervertebral disks become ballooned as they indent soft vertebral bodies, forming the "codfish spine."

3. *Pathologic Fractures.* These occur frequently.

4. *Marrow Fibrosis.* Replacement of marrow elements may cause anemia.

5. *Brown Tumors.* These are not actually tumors but are localized accumulations of hemorrhage and blood pigments and reactive masses of osteoclasts in a spindle cell stroma. The "tumor" is a well-circumscribed dark-brown area of soft consistency situated where bone resorption has been thorough. Healing may occur by fibrous tissue replacement, or the center may liquefy, and a bone cyst remains.

6. *Multiple Bone Cysts.* These are unilocular or multilocular; they expand the cortex, leaving a paper-thin covering, and are often the site of pathologic fractures. The walls are composed of dense fibrous tissue, and the contents are serous fluid and fibrin.

7. *Healing.* During the active resorptive stage of the disease, an attempt at replacement is observed in thin seams of osteoid apposition. Osteoid formation is especially pronounced at sites of stress, fractures and bending deformities.

After parathyroidectomy, osteoblasts become sparse, and osteoblastic activity becomes pronounced. Cortices thicken. Brown tumors disappear and are replaced by bone or become converted into cysts. The smaller cysts usu-



Fig. 28. Effect of hyperparathyroidism on bone. Marked osteoclastic resorption of trabeculae. Marrow spaces filled with vascular fibrous tissue.



FIG. 29. Renal hyperparathyroidism. Multiple areas of cystic bone absorption.

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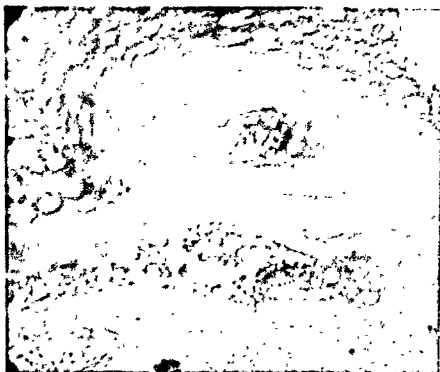
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FIG. 28. Effect of hyperparathyroidism on bone. Marked osteoclastic resorption of trabeculae. Marrow spaces filled with vascular fibrous tissue.

Plate 6. Osteomalacia. The wide pink-staining osteoid seams are deficient in calcium salts. (× 70)



Bone destruction exceeds bone formation. New protein matrix (pink-staining osteoid) is being laid down in a strong effort at reconstruction. This is reflected in the blood stream by an increase in the alkaline phosphatase level. However, the osteoid tissue remains uncalcified to a large degree, accumulates in large amounts and softens the bone (osteomalacia) which in turn yields to stresses and strains.

When this chain of events takes place during infancy and childhood, longitudinal growth is affected. The process of endochondral ossification at the epiphyseal growth plate demands that the cartilage be calcified before it can be resorbed and replaced by osteoid. The latter in turn is meagerly formed at this site and remains uncalcified.

Excessive mobilization of calcium from the skeleton, when not caused by deficient absorption from the intestine, is caused by hyperparathyroidism, primary or secondary to renal disease.

Rickets (Rachitis)

Rickets is defined as a disease of infancy and childhood due to insufficiency of calcium and characterized clinically chiefly by softened and deformed bones. The typical clinical condition to be described forthwith is extremely rare except under circumstances of famine. However, the orthopaedic surgeon should bear in mind that, regardless of present-day prophylactic vitamin and nutritional care, a low grade subclinical type is frequently responsible for certain deformities such as knock knee and bowleg. Often such a patient, when given additional amounts of vitamin D, will display surprising spontaneous correction. When florid rickets yields only to very high dosage of vitamin D, the case is classified as vitamin D resistant rickets. Apparently some unknown factor causes a variable *degree of response to the vitamin.*

Etiology. Calcium lack is due to vitamin D deficiency, intestinal diseases, and dietary lack of calcium and phosphorus.

Vitamin D promotes the absorption of these minerals from the intestine. It is a fat-soluble vitamin, usually associated in foods with vitamin A and found in strongest concentration in fish-liver oils. It is present in lesser amounts in milk, cream, butter, egg yolk and animal fats. It is prepared commercially by irradiation and ergosterol, a lipoid obtained from yeast. The ultraviolet rays of the sun or a mercury quartz lamp convert the sterols of the skin to vitamin D. Dust, window glass and skin pigmentation will impede penetration of ultraviolet rays.

Intestinal diseases such as steatorrhea, celiac disease and sprue as well as the common diarrheas cause their effect in one of three ways: (1) fat is inadequately digested, combines with calcium and phosphorus to form



PLATE 5. Osteitis fibrosa cystica (hyperparathyroidism). Active bone resorption by multinucleated giant cell osteoclasts. Little or no new bone formation. This same histologic picture may also be observed about the lesions of Paget's disease, fibrous dysplasia and the wall of a bone cyst during the stage of active local resorption. A generalized distribution of active osteoclastic resorption throughout the skeleton identifies this as hyperparathyroidism. (× 108)

tive Chvostek and Trousseau signs, and convulsions. Treatment is either by administering expensive parathormone, or better by dihydrotachysterol (A. T. 10) administered orally 2 to 4 cc. daily. Its dosage must be regulated daily by the Sulkowitch test, a slightly subnormal serum calcium being preferred. Calciferol (vitamin D_2) is used occasionally. It is cheaper but not as effective as A. T. 10. The diet must be high in calcium but low in phosphorus. Milk is contraindicated because of its high phosphorus content. Intravenous injections of calcium effectively relieve attacks of tetany.

PSEUDOHYPOPARATHYROIDISM

This rare disease first described by Albright is characterized by lack of normal response to parathormone, the production of which is adequate. It is identified by failure of the individual to demonstrate the typical clinical and blood chemistry changes following administration of parathyroid extract. Fortunately, the condition is improved by A. T. 10 and Calciferol.

LOW PHOSPHORUS RICKETS AND OSTEOMALACIA

This defect is caused by lack of absorption of phosphorus from foodstuffs due either to dietary insufficiency or vitamin deficiency. The calcium level in the blood stream is maintained by normal parathyroid activity.

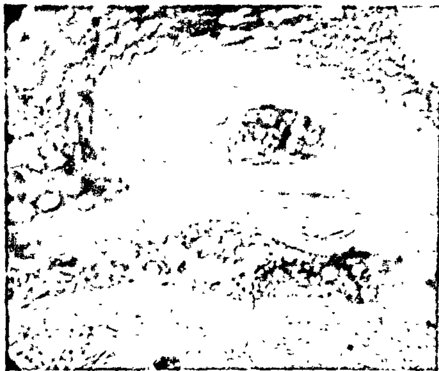
On the other hand, rickets and osteomalacia which shows a high serum phosphorus and low calcium is associated with compensatory parathyroid hyperplasia which excretes the excess of phosphorus and elevates the calcium level at the expense of the bone.

These conditions will be described fully under their respective headings.

CALCIUM-DEFICIENCY DISEASES

A physiologic level of serum calcium must be maintained or the state of hypocalcemia will result, characterized chiefly by muscle hyperirritability. Calcium is supplied from the diet and from bodily stores, mainly from bone. Within the bone, processes of new bone formation and old bone resorption are taking place constantly. When calcium absorbed from the gastro-intestinal tract is insufficient, the serum level is restored by mobilization from bone. The parathyroids undergo compensatory hyperplasia, and their effect is seen within the bone as ingrowth of vascular fibrous tissue replacing marrow and very active osteoclasis.

PLATE 6. Osteomalacia. The wide pink-staining osteoid seams are deficient in calcium salts. (× 70)



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Vitamin D promotes the absorption of these minerals from the intestine. It is a fat-soluble vitamin, usually associated in foods with vitamin A and found in strongest concentration in fish-liver oils. It is present in lesser amounts in milk, cream, butter, egg yolk and animal fats. It is prepared commercially by irradiation and ergosterol, a lipoid obtained from yeast. The ultraviolet rays of the sun or a mercury quartz lamp convert the sterols of the skin to vitamin D. Dust, window glass and skin pigmentation will impede penetration of ultraviolet rays.

Intestinal diseases such as steatorrhea, celiac disease and sprue as well as the common diarrheas cause their effect in one of three ways: (1) fat is inadequately digested, combines with calcium and phosphorus to form



FIG. 32. Rickets. (*Left*) Thickened and broadened epiphyseal plates. Ossification of the metaphysis is irregular and defective. The metaphysis is wide, its margin pointed, and the central area indented, resulting in a saucer-shaped appearance. The shaft is bowed, the cortex thin on the convex side, thick on the concave. (*Right*) Showing broad, saucer-shaped metaphyses, bowing of the diaphyses, and thickening of the cortex on the side of the concavity.

soaps which are eliminated; (2) vitamin D, being fat soluble, is likewise eliminated; (3) intestinal irritation reduces absorptive power

Insufficient calcium and phosphorus implies a diet consisting of foods with minimal amounts of these minerals. Milk, cheese, nuts and cabbage contain an abundance of calcium phosphate.

Rickets is rare during the first 6 months because of ample nutrition derived in utero. Prenatal calcium and vitamin lack in the mother (osteomalacia) is the main cause of *fetal rickets*. *Infantile rickets* is the most common type, developing between 6 months and 3 years. *Late rickets or rachitis tarda* is rare and is observed under conditions of famine or vitamin resistance (q.v.). An important complication at this age is separation of epiphyses, especially of the upper femoral epiphysis.

The disease occurs mainly in winter months and in nontropical regions, explained by deficient sunlight. The Negro and the Italian races exhibit a marked predisposition. A child who grows rapidly in height is more likely to develop rachitic changes than one whose growth is slow.

Pathology.[3] The histologic feature of rickets (and osteomalacia) is osteoid tissue which is the protein base in which the calcium and phosphorus salt has failed to deposit. Rickets and osteomalacia are fundamentally identical disturbances. Osteomalacia is the term applied after the epiphyseal plates have disappeared. The process may be divided into active and healing stages.

ACTIVE STAGE. At the *epiphyseal plate*, the orderly progression of endochondral ossification is interrupted. Proliferation of cartilage cells, palisade arrangement, and formation of matrix proceeds normally, but calcification of the matrix is deficient. This step is a necessary prerequisite to laying down of new bone about calcified cartilage. Capillary inshoots from the metaphysis invade and destroy the chondro-

[3] Luck, J. V. Bone and Joint Diseases, Springfield, Thomas, 1950.

cytes and lay down osteoid about large islands of noncalcified cartilage. The cartilage cell columns proliferate meanwhile to 10 to 20 times the normal depth of cells in a haphazard arrangement. The enormous accumulation of proliferated cartilage and osteoid tissue results in a widened, irregular epiphyseal line of radiolucency extending deeply into the metaphysis.

In the *metaphysis and the diaphysis*, thick layers of osteoid are laid down about thin old residual fragments of bone and within enlarged haversian canals. Pink-staining tissue appears everywhere, producing large bizarre-shaped, disorderly arranged trabeculae. A layer of osteoid often develops subperiosteally as an exuberant growth near the epiphyseal plate. Characteristic globular enlargements are formed in typical situations such as the costochondral junctions of ribs (rachitic rosary). The marrow displays a moderate degree of vascularity and fibrosis.

HEALING STAGE. Calcium salts are deposited in the *zone* of preparatory calcification. Capillaries penetrate the columns of proliferated chondrocytes and lay down osteoid about the calcified cartilage. The osteoid promptly becomes calcified and transformed to bone. The thickness of the epiphyseal plate is reduced to normal size. Osteoid trabeculae throughout the metaphysis and the shaft after conversion to bone resume their normal architecture. The fibrotic marrow is replaced by fatty and hematogenous elements. A bowed extremity often corrects itself spontaneously.

Symptoms. A history of dietary deficiency may be obtained. The infant displays increased restlessness at night, profuse diaphoresis, pallor of the skin, and a disinclination to play. A *generalized* catarrh of the mucous membranes is manifested by diarrhea, respiratory infection, etc. Occasionally, irritability of the central nervous system (hypocalcemia) results in spasmophilia, convulsions, Chvostek's sign, opisthotonus, etc.

Clinical Findings. The following are characteristic:

1. *Head:* large bosses over frontal and parietal eminences, flattening of occiput and vertex, causing enlarged squared appearance (caput quadratum). Fontanelles late in closing. Bones thin and crackling (craniotabes).

2. *Chest:* beading enlargements at costochondral junctions (rachitic rosary); a horizontal depression (Harrison's groove) a few inches above the lower costal margin is caused by the pull of the diaphragm on the softened ribs. The chest cage is narrowed transversely and elongated anteroposteriorly (pigeon breast).

3. *Abdomen:* prominent.

4. *Pelvis:* compressed transversely by weight-bearing, inlet is narrowed.

5. *Enlarged epiphyses:* especially at centers of most rapid growth (knees, wrist).

6. *Delayed dentition.*

7. *Skin pallor*, secondary anemia.

8. *Poor tone of muscles*, delayed walking.

9. *Deformities:* The lower extremities are most often deformed consequent to pressures of weight-bearing. Knock knees are usually due to deformity at the lower end of the femurs. Bowlegs are usually caused by lateral bowing and internal torsion of the tibiae. Coxa vara causes a waddling type of gait. Ligamentous laxity about deformed joints is typical.

10. *Incomplete fractures* are frequent and are a result of an insignificant trauma.

11. *Growth restriction* lasts only a comparatively short time so that stature is not affected. It is only in nonrecognized rickets that dwarfism is allowed to develop over a long period of time.

Roentgenographic Findings. In the acute stage, the ossification center in the epiphysis becomes poorly defined and smaller. The epiphyseal border of the metaphysis is cup-shaped, ill-defined and frayed. These findings are most pronounced about epiphyseal plates exhibiting the greatest rate of growth, e.g., lower end of femur, upper end of tibia. The metaphyseal cortices flare outward (trumpeting) and the epiphyseometaphyseal junction is widened. There is reduction in the secondary transverse trabeculae; the longitudinal trabeculae persist but are quite thin. The cortices are less sharply defined (because of osteoporosis and osteoid deposition). The bones, especially the long weight-bearing bones, bend, the cortices thickening on the concave side.

In healing, a dense line appears at the epiphyseometaphyseal juncture. This is the newly calcified cartilage. The epiphyseal line becomes narrower and well *defined*. The epiphyseal ossified nucleus becomes more

dense, larger and well defined. Transverse trabeculations reappear, and cortices resume their density and definition. Bending deformities with continued growth often disappear or subside to a great degree.

Laboratory Findings. The serum phosphorus is typically reduced. Often the serum alkaline phosphatase is high. Compensatory hyperparathyroidism generally maintains a normal serum calcium but at the same time effects excretion of phosphorus. When this mechanism is defective, serum calcium falls, and symptoms of neuromuscular irritability ensue.

Treatment. Prophylaxis consists chiefly of administration of vitamin D and exposure to sunlight, especially for premature infants and those on artificial milk feedings. Active treatment by vitamin D, calcium preparations and ultraviolet rays will effect healing demonstrable on roentgenograms within 2 weeks. Deformity of the lower extremities usually spontaneously regresses to a great degree over a period of months. The application of braces or osteotomy to correct deformity is generally unnecessary. Failure of spontaneous correction usually signifies an inadequate dosage of vitamin D. Rarely, when deformity persists, osteotomy is indicated but is best postponed until growth is complete. Knock knee and bowleg are discussed in detail in the section on the Knee.

The older texts describe *osteoclasis*, the procedure of manually fracturing a deformed bone by force over a wedge or by use of a Thomas osteoclast, a wrenchlike device. The author deprecates this method as unnecessarily traumatic and possibly endangering epiphyseal growth.

Celiac Rickets

Idiopathic steatorrhea, also known as celiac disease, nontropical sprue, Gee's disease or Gee-Thaysen disease, is a condition characterized by soapy stools, in which calcium and phosphorus are lost. A deficiency of the pancreatic lipolytic enzyme causes incomplete digestion of fats which therefore cannot be absorbed; consequently, vitamin D is not absorbed. An excess of free fatty acids combines with calcium and phosphorus, forming precipitates of soap which are excreted as such in the stool. Calcium and vitamin D loss results in the picture of rickets.

Treatment consists of suitable dieting, administration of high potency preparations of vitamin D and calcium lactate intramuscularly, and ultraviolet treatment.

Renal Causes of Rickets and Osteomalacia

(Renal Dwarfism; Renal Pseudorickets; Renal Osteitis Fibrosa Cystica)

Renal insufficiency, whether a result of glomerular or tubular disease, is associated with compensatory parathyroid hyperplasia which deprives the skeleton of calcium. Typical hyperparathyroid histologic changes in the bone consist of active osteoclastic resorption and fibrosis, the calcium being mobilized to combat the renal acidosis. Failure of absorption of calcium and vitamin D from the intestine does not occur so that uncalcified osteoid tissue does not form as extensively as it does in rickets and osteomalacia. However, even the dietary calcium becomes relatively unavailable for bone so that a slight degree of osteoid and bony softening develops. Therefore, weight-bearing deformities are not as pronounced as in rickets.

When the renal factor is present at birth, it is due to congenital cystic disease or congenital hydronephrosis. Later in life chronic glomerulonephritis, chronic interstitial nephritis, and the nephroses due to heavy metal poisoning are etiologic factors. Congenital kidney conditions have an insidious slowly progressive effect throughout childhood, usually becoming clinically manifest at about puberty or adolescence. Typical rachitic changes chronically interfere with endochondral ossification at the epiphyseal plates, thereby restricting longitudinal growth. Shortness of stature and enlarged epiphyses are characteristic. True florid rickets is never seen. The condition is simply low-grade rickets, because of unavailable calcium, superimposed upon osteitis fibrosa.

The ensuing classification follows the concepts advanced by Albright and his associates (Renal Osteitis Fibrosa Generalisata):

1. Renal Disease With Phosphate Retention. The retention of nonprotein nitrogen is almost always associated with retention of phosphorus. The serum calcium level becomes lowered because *(1) it adjusts to the high serum phos-*

phorus level, and (2) it is used up as a base to combat the acidosis. The parathyroids hypertrophy to excrete more hormone which acts either (1) by promoting excretion of serum phosphates, or (2) by promoting resorption of bone tissue. Pathologically, one sees a predominance of bone destruction rather than a delay in the calcification of newly formed osteoid.

Clinically, in children before union of the epiphyses, in addition to bony changes of osteitis fibrosa cystica, changes are seen at the epiphyseal line almost identical with those of rickets. Slipping of the upper femoral epiphyses is not uncommon. Retardation of longitudinal growth is a constant finding.

The characteristics of renal osteitis fibrosa generalisata in adults include:

A. Renal insufficiency which is longstanding and severe

B. Nitrogen and phosphorus retention

C. Normal or slightly low serum calcium level

D. Severe acidosis with a low CO_2 combining power of serum

E. High serum chloride or low serum sodium level

F. Arteriosclerosis of the Monckeberg type (*medial arteriosclerosis*)

G. High serum phosphatase level

H. Sometimes calcium deposits around joints

I. Roentgenograms display a generalized decalcification with a cystic or motheaten appearance

2. Renal Disease With Excessive Phosphorus Loss. Two separate conditions can be distinguished in which failure of tubular function results in excessive phosphate excretion:

A. *Failure of Tubules to Form a Base With Which to Excrete Acid in Urine* ("tubular-insufficiency-without-glomerular-insufficiency" of Albright, et al.). As demand is made upon calcium which appears in increased amounts in the urine, the low serum calcium level leads to a low serum phosphorus level. Low serum calcium and phosphorus causes failure of mineral deposition in bone. Osteomalacia develops, is less resistant to stresses and strains, osteoblasts are stimulated, and a high serum alkaline phosphatase results. The low serum phosphorus delays calcification of newly formed osteoid so that a greater amount of this tissue is seen.

Hypopotassemia is a common complication of renal acidosis. Potassium, like calcium, is utilized as a base and is excreted in excess in the urine. Symptoms of the low potassium syndrome include pain in the extremities, inability to move the arms and the legs, and electrocardiographic changes.

Nephrocalcinosis and nephrolithiasis frequently accompany this form of renal acidosis. Alkali therapy decreases calcium excretion in the urine and lessens formation of new stones.

The etiology of tubular-insufficiency-without-glomerular-insufficiency is obscure.

B. *Fanconi's Syndrome.*[4] In this syndrome, the renal tubules fail to absorb phosphates, glucose and many of the amino acids. Consequently, a *low serum phosphorus* is associated with *glycosuria* and *aminoaciduria*.

Fanconi in 1936 described the condition as characterized by a hereditary tendency, often a history of consanguinity, retarded growth, rickets, albuminuria, glycosuria, persistently alkaline urine, increase of organic acids, ammonia, phosphorus, and calcium in the urine, marked hypophosphatemia without hypercalcemia, lowering of the blood bicarbonate *without azotemia*, and degenerative changes in the renal tubular epithelium. Cystine deposits in almost all organs, mainly the reticuloendothelial system, may be associated if the kidney is unable to excrete cystine.

The mechanism of production of late rickets and osteomalacia seems to be an acidosis due to increased urinary excretion of base secondary to increased urinary excretion of organic acids.

Symptoms of this disorder appear early in childhood and become progressively worse.

Treatment of Renal Rickets and Osteomalacia. Properly treated, the relief of skeletal symptoms is often spectacular. Alkaline salts are administered, especially those combinations of the salt of a mineral base with an organic acid, e.g., sodium citrate, sodium lactate, or calcium gluconate. If hypopotassemia is a factor, potassium citrate is added. The organic acid is destroyed, leaving the base free

[4] McCune, D. J., Mason, H. H., and Clarke, H. T.: Intractable hypophosphatemic rickets with renal glycosuria and acidosis (The Fanconi Syndrome), Am J. Dis. Child. 65:81, 1943.

to help in the excretion of acid in the urine. Citric acid may be given to increase gastric acidity and aid calcium absorption. To overcome the osteomalacia, a high calcium intake and massive doses of vitamin D will effect a strongly positive calcium balance. Osteomalacia is cured, and normal growth is resumed. Thereafter, loss of calcium can be prevented by alkali therapy alone. Continuation of vitamin D in massive doses is not advisable for fear of causing vitamin D poisoning.

Vitamin Resistant Rickets[5, 6] (Refractory Rickets)

Formerly thought to be rare, refractory rickets is now known to be quite common and probably is the most frequent cause of dwarfism. Compared with the usual form of rickets, refractory rickets is more severe, fails to respond to usual doses, but responds to massive doses of vitamin D. Albright's metabolic studies revealed that *vitamin D is absorbed but the patient does not respond until a threshold level has been exceeded.* An adequate response is shown by increased urinary excretion of calcium and decreased fecal excretion of calcium. The serum phosphorus rises, and the alkaline phosphatase subsides to normal. Roentgenograms reveal healing at the epiphyseal lines, and growth is resumed at the rate of 1 cm per month.

Clinical Picture. A marked familial tendency is often observed. The patient is of short stature with all the usual signs of florid rickets. Deformities are severe, particularly in the lower extremities, where bowlegs, knock knees and a "tackle deformity," consisting of a bowleg on one side and a knock knee on the other, are seen. Marked ligamentous instability is typical A waddling gait is due to coxa vara These deformities are persistent and typically recur after attempted osteotomies. The skull has a characteristic appearance. The anteroposterior diameter is increased, and the transverse diameter is decreased (dolichocephaly); frontal bossing and a marked external occipital protuberance occur. The nose is often saddle-shaped.

[5] Albright, F, Butler, A. M, and Bloomberg, E Rickets resistant to vitamin D therapy, Am J. Dis Child. 54:629, 1937

[6] Pederson, H. E, and McCarroll, H. R.: Vitamin resistant rickets, J Bone & Joint Surg 33A:203, 1951

Roentgenographic Findings. These are the usual findings of rickets. However, the trabeculae are coarser, broader and more widely spread than usual.

Laboratory Findings. The serum phosphorus is generally below 3 mg., the calcium is normal and the alkaline phosphatase is elevated to 20 or more Bodansky units.

The urinary qualitative Sulkowitch test is negative or reveals only a trace of calcium. The urine concentration is normal, and its reaction is acid.

Differential Diagnosis. Cases which in the past have been wrongly classified as achondroplasia, dyschondroplasia and chondrodysplasia, (and their hopeless prognosis) are now properly identified by blood and urine studies plus response to high dosage of vitamin D.

The condition must also be differentiated from renal rickets and the Fanconi syndrome by its decreased urinary calcium excretion, absence of glucose, albumin and amino acids in the urine, and absence of disturbed plasma electrolytes.

Picture in Adults. This is characterized chiefly by dwarfism. The alkaline phosphatase is usually normal, but variable phosphorus levels are observed. The roentgenogram displays a coarse architecture of trabeculae, Looser lines, vertebrae typically biconcave with ballooned disks, degenerative changes at the knees, the hips and the lumbosacral spine. At the points of attachment of large muscle masses, a bony protuberance suggests the effect of pronged traction upon soft bone. Various bony deformities, especially about the knees, have existed since childhood.

Treatment. The aim of treatment is to provide a high dosage of vitamin D and maintain the Sulkowitch at 1 to 2+ (at this level urinary calcium casts do not appear). First, the threshold level is determined by increasing the dosage until the Sulkowitch shows that adequate urinary excretion of calcium is occurring. Then the dosage is increased until the serum phosphorus level reaches 5 mg., or toxic symptoms appear. These toxic symptoms include anorexia, nausea, vomiting, weight loss, occasional hematuria, rarely oliguria or anuria with nephrocalcinosis. Toxicity is not seen when the serum calcium is kept below 12 mg. and the Sulkowitch below 3+. Above 12 mg.,

nephrocalcinosis occurs, about 16 mg. general metastatic calcification develops. Blood and urinary changes are noted within a week and x-ray changes are observed after 3 to 4 weeks. Vitamin D must be continued until growth is complete to ensure adequate continuance of growth. After cessation of growth persistence of serum and urinary findings indicates that osteomalacia is present, and then vitamin therapy must be continued.

Attempted surgery, especially osteotomies for correction of deformities, are best delayed until the epiphyses have closed. Otherwise recurrence is almost inevitable. When osteotomy is performed and followed by plaster immobilization, hypercalcemia and hypercalciuria develop, and the extremely high concentration of calcium in the urine may interfere with renal function. Therefore preoperatively it is advisable to suspend vitamin D administration until 2 weeks postoperatively.

In adults with demineralization and deformity of the spine, especially in a subject of short stature, consideration must be given to the possibility of vitamin D resistance. Appropriate metabolic study may reveal this condition.

Osteomalacia
(Mollities Ossium)

Osteomalacia is a condition characterized etiologically by calcium and vitamin D deficiency in the adult, and pathologically by softening of the bones due to an excess of uncalcified osteoid tissue. Therefore, it is identical with rickets of infancy and childhood except that longitudinal growth is unaffected.

Etiology. The process of catabolism (osteoclastic resorption) in bones throughout the body continues normally. On the other hand, the process of anabolism continues normally only as far as the laying down of the protein matrix (osteoid), but the hardening precipitation of lime salts does not occur for lack of calcium. Insufficiency of calcium is due either to inadequate intestinal absorption or excessive loss:

A. Failure of Intestinal Absorption

1. *Dietary lack of calcium*, e.g., famine.
2. *Insufficient vitamin D*, necessary to absorption of calcium and deposition in bone, e.g., lack of sunshine. The condition is most common in India where the Purdah custom demands that women keep their faces and bodies covered and remain indoors.
3. *Gastro-intestinal disorders*, as sprue or steatorrhea. Diarrhea causes rapid passage of intestinal contents, thereby reducing calcium absorption.
4. *Laxatives*. Mineral oil dissolves vitamin D and mineral salts form insoluble calcium compounds.

B. Excessive Loss

1. *Pregnancy*, in the last 3 months when demands are made by development of the fetal skeleton.
2. *Lactation*, which makes an intense demand upon the skeleton when dietary calcium is inadequate. The daily loss may be as high as 250 to 350 mg.
3. *Renal Acidosis*. An excessive amount of acid radicals must be neutralized because of impaired efficiency of the kidney to excrete. The increased demand for neutralizing bases uses up the available calcium, which leaves an inadequate supply for calcification of osteoid. (See section on Renal Causes of Rickets and Osteomalacia.)
4. *Idiopathic Hypercalciuria*. A low renal threshold for calcium permits a continuous excretion of a large amount of calcium, thereby depleting the skeleton.

Pathology. The basic finding is an excess of persisting osteoid seams, which surround thin, old trabeculae. Here and there a normal degree of osteoclasis is proceeding. Much of the compact bone is transformed into cancellous bone. Osteoblastic activity continues, and layer upon layer of osteoid tissue is formed. The development of osteoid is most pronounced at sites of maximal stresses and strains. The marrow appears to be vascular and fibrous.

Fractures are usually multiple and heal with an abundant callus formation consisting *chiefly of osteoid. Grotesque deformities* develop, due to bending with weight-bearing pressures.

Clinical Picture. *Deformities*, particularly of weight-bearing structures constitute the chief finding. The leg and the thigh are se-

to help in the excretion of acid in the urine. Citric acid may be given to increase gastric acidity and aid calcium absorption. To overcome the osteomalacia, a high calcium intake and massive doses of vitamin D will effect a strongly positive calcium balance. Osteomalacia is cured, and normal growth is resumed. Thereafter, loss of calcium can be prevented by alkali therapy alone. Continuation of vitamin D in massive doses is not advisable for fear of causing vitamin D poisoning.

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Pathology. The basic finding is an excess of persisting osteoid seams, which surround thin, old trabeculae. Here and there a normal degree of osteoclasis is proceeding. Much of the compact bone is transformed into cancellous bone. Osteoblastic activity continues, and layer upon layer of osteoid tissue is formed. The development of osteoid is most pronounced at sites of maximal stresses and strains. The marrow appears to be vascular and fibrous.

Fractures are usually multiple and heal with an abundant callus formation consisting chiefly of osteoid. Grotesque deformities develop, due to bending with weight-bearing pressures.

Clinical Picture. *Deformities,* particularly of weight-bearing structures constitute the chief finding. The leg and the thigh are se-

verely bent. Scoliotic and kyphotic deformities of the spine develop. Pressure of the femoral heads indent the acetabulae and the lateral walls of the pelvis.

Generalized skeletal pain and tenderness occur but may be confined to the lower back and the lower extremities. An acute onset of localized pain and tenderness may signify an incomplete fracture. Muscle weakness is typical.

Other symptoms are related to the causative dietary, gastro-intestinal, pregnancy, or renal factors

Laboratory Findings. The serum findings are a normal or low calcium, a low or normal phosphorus (most commonly a normal serum calcium with a low serum phosphorus), and a high alkaline phosphatase.

Roentgenographic Findings. Osteomalacia effects a generalized demineralization with loss of transverse trabeculae not unlike the appearance of osteoporosis. Laboratory and clinical findings may be necessary to distinguish the two conditions. It is also important to recognize that osteomalacia may be present without x-ray evidence.

The two diagnostic findings when bone changes are pronounced are *demineralization* and persistent transverse *Looser zones*. The skeleton is diffusely rarefied, and the cortices are thinned. There is no subperiosteal resorption of bone in contrast with hyperparathyroidism. Thus the lamina dura around the teeth is present in osteomalacia and absent in hyperparathyroidism.

Looser lines or pseudofractures are frequently found. These are transverse bilaterally symmetrical lines of rarefaction which extend incompletely across the bones. Albright believes that these represent incomplete fractures which have healed by callus which consists of osteoid tissue, persisting for lack of calcium. A Looser line often occurs in a bone which otherwise may appear to be normal and as a matter of fact may be the only evidence of osteomalacia. It occurs repeatedly at definite points, the necks of the femurs, the rami of pubic and ischial bones, ribs, and typically in the axillary edge of the scapula. It lasts for months or years without regressing, is bilaterally symmetrical and occurs only in rickets and osteomalacia. Occasionally, the periosteum overlying these areas may be seen as elevated with a slight deposit of subperiosteal bone. These osteoid zones invariably heal when the cause of the osteomalacia is identified and appropriate treatment is given.

Similar x-ray appearances of fractures in other diseases are really a fibrous and cartilaginous tissue of delayed union. In Paget's disease, polyostotic fibrous dysplasia and osteogenesis imperfecta, these fractures typically occur at the site of localized bone pathology and do not respond to measures used to overcome osteomalacia.

As a result of bone softening, bending deformities are seen. In the spine, the vertebral changes are common to all bone conditions leading to demineralized vertebrae. The nucleus pulposus expands the disks and indents the end plate of the vertebral body. All bodies become biconcave, and the typical "codfish" spine results. Compression fractures often occur.

Differential Diagnosis

1. *Osteoporosis.* This is a disturbance of tissue metabolism, not calcium metabolism. Not enough matrix is laid down by the osteoblasts, but whatever is formed is calcified. Therefore, calcium, phosphorus and alkaline phosphatase levels are normal. Biopsy reveals absence of osteoid tissue.

2. *Osteitis Fibrosa Generalisata.* Bone tissue decreases because of increased bone resorption. The compensatory increased activity of osteoblasts leads to a high alkaline phosphatase. The commonest cause is hyperparathyrodism which causes low serum phosphorus and high serum calcium levels. The lamina dura is absent. Cystic or motheaten changes in the bone are often seen.

Treatment. It must be remembered that low-grade degrees of osteomalacia are quite common. The demands of the skeleton may be just barely met until some supervening condition such as pregnancy deprives the body of calcium. Generalized bone pains and tenderness may be the only clue. Or a compression fracture of a vertebral body or a Looser line may be observed. Blood serum values may be normal. Where a low grade type of osteomalacia is suspected, a therapeutic trial is warranted.

Calcium in the form of lactate or gluconate

(0.5 to 3.0 Gm.) is given 3 times a day. Dicalcium phosphate is also useful.

About 10,000 units of vitamin D is administered daily, and as healing takes place the dosage is reduced to 400 units for children and 500 units for adults. The dosage of vitamin D when treating active rickets and osteomalacia must be adequate but not excessive.

The diet must be high in protein. The minimum requirement is 3.5 Gm. per Kg. for infants down to 1.0 Gm. per Kg. for adults. Animal protein will supply substantial amounts of phosphorus. Meat, seafood and dairy products are excellent sources of phosphorus, calcium and protein.

Gastro-intestinal disorders which interfere with calcium absorption must be corrected. Achlorhydria requires administration of dilute acids with each meal. Bile salts aid digestion of fat which otherwise would combine with and prevent calcium absorption. Other substances which affect absorption of calcium, phosphorus or vitamin D must be reduced or avoided. Examples are:

Mineral oil prevents vitamin D absorption.

Phytins decrease calcium available for absorption.

Oxalic acid combines with calcium and prevents its utilization even after absorption

Aluminum hydroxide gel prevents phosphorus absorption.

Renal conditions require alkalinizing agents in addition to calcium and vitamin D. Once symptoms of osteomalacia have been overcome, only alkali need be continued. (See Renal Causes of Osteomalacia and Rickets.)

Correction of deformities is accomplished by osteotomies.

MALABSORPTION SYNDROME[7, 8]

This is the term applied to the clinical picture produced by a wide variety of conditions which reduce the absorptive ability of the small intestine The main defect appears to be failure to absorb fat and fat-soluble vitamin D, glucose, vitamin A, vitamin B_{12} and the intrinsic factor.

[7] Bossak, E. T., Wang, C I., and Adlersberg, D.. Clinical aspects of the malabsorption syndrome, J. Mt Sinai Hosp 24:286, 1957

[8] Hartley, J . Osseous changes and fractures in the malabsorption syndrome, J. Mt. Sinai Hosp. 24 346, 1957

The dietary calcium combines with fatty acids to form insoluble soaps which with the excess of fats and carbohydrates are excreted in bulky, frothy, foul-smelling stools. In infants and children, the condition is called *celiac disease* and in adults it is designated as *idiopathic steatorrhea.* Deficiency of vitamin D and calcium is reflected in rickets and osteomalacia.

Deficiency of vitamin B_{12} and the intrinsic factor result in a macrocytic anemia and a beefy red tongue of glossitis. It is similar to pernicious anemia causing paresthesias, but achlorhydria and subacute combined degeneration of the cord is rare. A diffuse brown pigmentation is often observed over the exposed areas of the trunk and the extremities. This disturbance is designated *idiopathic sprue* when no pathology is demonstrable in the bowel, and *secondary sprue* when specific disease of the small bowel can be identified.

Also included in the malabsorption syndrome are a host of conditions such as *biliary obstruction* (bile necessary for emulsification of fat), *pancreatitis* (absence of lipolytic enzyme), and surgical conditions (gastrectomy, gastrojejunocolic fistula, etc.) which effect rapid propulsion of intestinal contents.

Clinical Picture. The malabsorption syndrome is characterized, in severe cases, by prolonged duration of symptoms including intermittent diarrhea, marked weakness, weight loss and glossitis. The patient appears poorly nourished and pale, the abdomen is distended, and often, because of a hypoproteinemia, dependent edema develops.

Osteomalacia, due to lack of vitamin D and calcium, is low grade and manifest by subtle findings such as slight bowing of the extremities and perhaps a Looser line or pseudofracture. In infants, rickets is produced. Osteoporosis is generalized and is a result of protein deficiency. Malnutrition will cause atrophy of the anterior lobe of the pituitary, and likewise the reduced trophic stimulation on sex hormone production will produce osteoporosis. Osteomalacia and osteoporosis frequently coexist in the same patient. "Bone pains" are common complaints, and pathologic fractures occur.

Severe cases of the malabsorption syndrome are uncommon. On the other hand, reduction of intestinal absorption in a subclinical state

is very common, particularly after abdominal surgical procedures. One usually does not observe typical diarrhea or a pronounced glossitis. Investigation of the causes of osteomalacia and osteoporosis should include attention to the malabsorption syndrome. The following laboratory procedures are essential to the diagnosis.

Laboratory Findings. The defect in absorption may affect one or several elements:

1. Macrocytic anemia
2. Hypoproteinemia
3. Hypocalcemia. In low-grade states, the compensatory hyperparathyroidism sustains the serum calcium level which therefore appears normal. A negative qualitative Sulkowitch test indicates a failure of calcium absorption.
4. Hypolipidemia, especially serum total and esterified cholesterol
5. Steatorrhea (may be present without diarrhea). The fecal fat exceeds the normal 20 per cent by dry weight.
6. A flat glucose tolerance curve when the glucose is administered orally; a normal curve if administered intravenously
7. Oral vitamin A curve low; intravenous curve normal.
8. Impaired absorption of radioactive vitamin B_{12}
9. Occasionally, a "typical" appearance of small intestine in roentgenograms

Treatment. Intramuscular administration of liver extract, folic acid and vitamin B_{12} is very effective in inducing a remission. A high-vitamin, high-protein diet, low in fat is prescribed. Until the absorptive defect is overcome, intramuscular calcium lactate, ultraviolet and the sex hormones are necessary for normal bone formation. This treatment must be continued indefinitely in the presence of permanent gastro-intestinal pathology.

HYPOPHOSPHATASIA[9, 10]

Hypophosphatasia is a condition clinically resembling rickets but typified by a persistently low serum alkaline phosphatase. It is probably hereditary.

Clinical Picture. The following findings are characteristic:

1. *Stormy infancy:* failure of weight gain, periodic attacks of vomiting, delay in walking, delayed dentition
2. *Stunted growth:* shortness of long bones
3. *Rachitic bony changes:* deformities, thickening about epiphyses, beaded ribs, etc.
4. *Liability to fractures*, especially at metaphyses; healing is not delayed and takes place in normal fashion.
5. *Premature loss of deciduous teeth*
6. *Craniostenosis* may cause impaired vision; the skull is peculiarly soft and leather like.
7. *Gait unsteady*, poor tolerance to exercise

Laboratory Findings

1. *Roentgenograms:* generalized demineralization, rachitic irregularities about epiphyseal lines
2. *Low serum alkaline phosphatase*
3. *Excess urinary excretion of phosphoethanolamine*
4. *Hypercalcemia and hyperphosphatemia.* May be normal.

Types of Cases. *Severe* cases are usually fatal during the first year of infancy, usually attributable to a renal lesion. If the infant survives this period, the outlook for recovery is usually good, the condition remaining stationary or improving and becoming the *moderately severe* cases. *Mild* cases are those without bone changes.

SCURVY (Scorbutus)

Scurvy is a nutritional disorder caused by deficiency of vitamin C and characterized clinically by a generalized hemorrhagic tendency. The severe form of the disease is rare, but mild and subclinical types are relatively common. Its main effects are on cells and tissues of mesodermal origin, particularly in the skeletal system.

Etiology. The disease occurs most frequently in artificially fed infants between the ages of 5 and 10 months. Since vitamin C is destroyed by heat, exclusive feeding with processed milk which is lacking in this vitamin will result in latent or symptomatic scurvy unless cevitamic acid or orange, lemon, or tomato juice is supplied. An infant fed ex-

[9] Rathbun, J. C.: Hypophosphatasia, Am J. Dis. Child 75:822, 1948.

[10] Dickson, W., and Harrocks, R. H.: Hypophosphatasia, J. Bone & Joint Surg. 40B:64, 1958.

Barton's Disease

clusively on milk necessarily is also deprived of vitamin D so that rickets also develops. The combination of rickets and scurvy is known as *Barlow's disease.*

Adult scurvy commonly occurs in elderly individuals who live on a restricted diet. Ordinarily, this vitamin C deficiency is subclinical and manifest by subcutaneous hemorrhages with slight trauma and delay in healing of wounds.

Pathology.[11] Vitamin C deficiency impairs the cohesive property of the matrix of connective tissue and endothelium. Consequently, capillary hemorrhages occur beneath mucous membranes and other locations of abundant capillary accumulations. Extraskeletal sites include the gums, intestines, conjunctivae, skin, bladder and kidneys. The most vascular skeletal situations are located beneath the periosteum and in the marrow, particularly in the metaphyses and especially adjacent to the most actively growing epiphyses (lower end of femur, upper end of tibia, upper end of humerus).

1. *Periosteum.* Subperiosteal hemorrhage is characteristic. The accumulation of blood may be slight or so extensive as to balloon out the periosteum and to resemble a large tumor. The clotted blood is either resorbed or transformed to fibrous tissue. Subsequently, especially when vitamin C is supplied, the organized hematoma becomes ossified with fine periosteal trabeculations. Eventually, with healing, the periosteal bone is resorbed.

2. *Epiphyses and Metaphyses.* Hemorrhages within the metaphysis interfere with ingrowth of osteoblastic tissue. Therefore, enchondral ossification proceeds normally only as far as formation of calcified cartilage (zone of provisory calcification) which accumulates in large amounts. Osteoblasts and osteoclasts are notably deficient or lacking. The broadened layer of calcified cartilage is known as *the white line of Fraenkel,* which appears as a characteristic transverse line of density on roentgenograms. When scurvy is less severe, a few irregular bone trabeculae may form, within which are contained unresorbed islands of calcified cartilage.

Within the epiphysis itself, a zone of calcified cartilage accumulates about the bony centrum. This encircling dense ring is known as *Wimberger's line.*

The metaphysis in response to hemorrhage becomes extremely hyperemic. The resultant resorption of bone in addition to failure of laying down of new bone results in extremely deficient ossification which appears in roentgenograms as a dark zone of radiolucency adjacent to the white line. Lack of bone structure and accumulation of fragile calcified cartilage weakens the epiphyseometaphyseal junction and leads to fractures and epiphyseal separation. The epiphysis and the attached epiphyseal plate may be completely displaced from the shaft. Nevertheless, with vitamin C treatment, although union occurs in the displaced position, continued growth restores normal contour to the bone.

3. *Marrow.* Hemorrhages throughout the marrow result in fibrous organization and replacement of hematopoietic tissue. Secondary anemia results.

4. *Bone.* Osteogenesis is interfered with, and osteoclasis continues. Trabeculations become thinned and poorly visualized in roentgenograms (ground-glass appearance). Cortices become slender and resemble those seen in osteogenesis imperfecta. Pathologic fractures occur through the metaphysis in infants and the diaphysis in adults.

5. *Teeth.* Porosis of alveolar bone permits loosening of the teeth. Pulp hemorrhages are followed by degeneration and necrosis. Dentine formation ceases, but the enamel is not affected. The gums are swollen and hemorrhagic.

Clinical Picture. The infant is restless, pale and febrile. The extremities are held immobile, the muscles are in spasms, and attempts to move them causes him to cry out with pain. A palpable, excruciating, tender, fixed swelling detected over a bone is the result of subperiosteal hemorrhage. If hemorrhage is recent, the swelling is soft and fluctuant. Later, it is indurated and less tender. Hemorrhages are particularly prone to develop above or below the knee. The voluntary immobilization of the extremities is termed *pseudoparalysis.*

The gums display a bluish, spongy swelling, especially about the upper central incisor teeth. The teeth are loose and brittle. Petechiae or ecchymoses are found in the skin or the mucous membranes. Hematemesis and

[11] Luck, J. V.: Bone and Joint Diseases, Springfield, Ill., Thomas, 1950



Fig. 35. Severe gouty arthritis of hands.

dral bone is replaced in well-circumscribed punched-out areas by the crystalline deposits. A pannus of granulation tissue grows over the articular surface, invades and replaces the cartilage and may bridge the joint to the opposite articular surface, producing a fibrous ankylosis. The irregularity of the joint surfaces leads to a secondary degenerative arthritis. Urate salts are deposited in the synovial membrane, the periarticular soft tissues and the subcutaneous tissues.

Microscopically the typical urate crystals are demonstrated by special technic. The deposits are surrounded by an inflammatory reaction, fibrous tissue and giant cells. The salts are found in articular cartilage, bone marrow, synovial membranes, joint capsules, ligaments, periosteum, tendons, bursae, subcutaneous and intramuscular tissues. The metatarsophalangeal joint of the big toe is predisposed. Next most affected are the intertarsal joints, the ankles, the fingers and the wrists. In the kidneys, dots of urate crystals are spread throughout the cortex and linear streaks through the medulla. A glomerular fibrosis is frequent. The cause of death is usually coronary or cerebral vascular disease, or nephrosclerosis with uremia.

Clinical Picture

The gouty patient definitely has a *hyperuricemia* for a number of years without symptoms. Measured in terms of urate salt, the serum urate invariably *exceeds 6 mg. per 100 cc.* The initial acute attack may come without warning and frequently is preceded by a *provocative factor* such as *trauma* (long walks), *dietary indiscretions* (high fat diet), *drugs* (liver extract), *surgical operations, exposure to cold* and *withdrawal of ACTH.* The patient is usually a man over 30. The attack has a sudden onset, frequently at night. The joint, often the metatarsophalangeal joint of the great toe, becomes very swollen, red and tender. The swelling is extreme, simulates a cellulitis, and extends beyond the confines of the joint. A variable amount of increased joint effusion contributes to the swelling. The joint fluid is particularly increased when the knee is the site of the attack. *Pain* is excruciating. The inflammation may involve a nonarticular urate deposit such as a subcutaneous tophus or in a bursa. *Constitutional symptoms* include fever, tachycardia, headaches, etc.

During the acute attack laboratory findings reveal a leukocytosis and an elevated sedimentation rate. The attack may last from a few days to several weeks before it subsides with complete restoration of function of the joint. *Desquamation* over the involved area is the final stage.

The first *interval* until the second attack is generally asymptomatic, and its duration is variable. Occasionally, several years may elapse before the next attack appears. Although the intervals vary in length, they tend

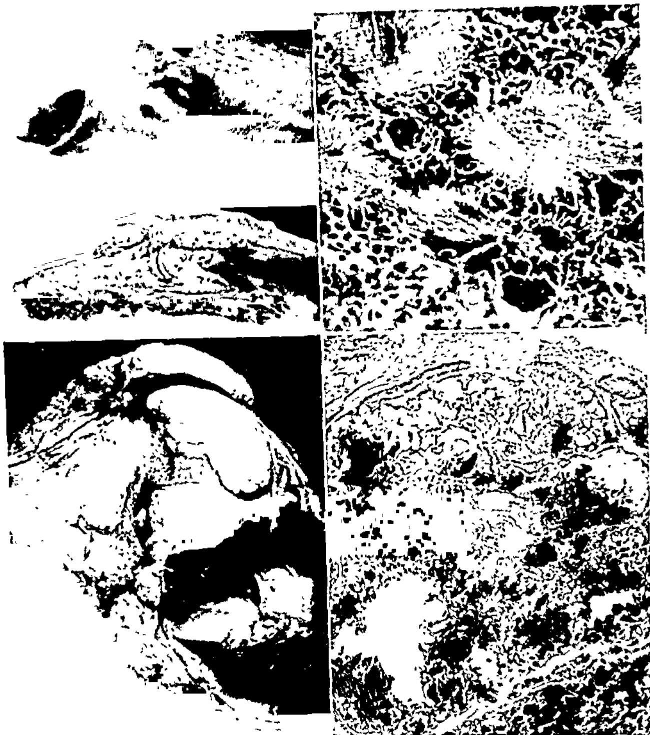


FIG. 36. Gout. (*Top, left*) The white crystalline deposits have invaded the phalanges, the tendon sheath and the tendon and have penetrated the skin. (*Bottom, left*) Extensive white urate deposits throughout a large joint. (*Top, right*) Microscopic appearance of a gouty nodule. The tissue has been fixed in absolute alcohol, because watery fixatives dissolve out the crystals. Note necrotic amorphous material, bundlelike accumulations of crystals, and reactive inflammatory cells including giant cells. (*Bottom, right*) A silver stain (Galanth's stain) which selectively darkens urate deposits. (Dr. Edward Rosenberg)

to become progressively shorter, and the intensity of succeeding attacks become progressively more severe. The later attacks are more likely to be febrile.

Small deposits of urate salts in the subcutaneous tissues are in evidence in the forearm where they have a pearly white appearance. A characteristic location for these deposits is



FIG. 37. Alkaptonuric (ochronotic) arthritis. The intervertebral disks are narrowed and calcified. (Harrold, A. J.: Alkaptonuric arthritis, J. Bone & Joint Surg. 38B:532)



prevention of joint destruction by immobilization. Large tophi which interfere with joint and tendon motion may be removed.

OCHRONOTIC ARTHRITIS

(Alkaptonuric Arthritis)[15]

Homogentisic acid is an aromatic acid resulting from the incomplete breakdown of the amino acids tyrosine and phenylalanine in the body. It has an affinity for certain tissues, particularly cartilage, which in consequence becomes brittle and disintegrates. A secondary degenerative arthritis ensues.

Etiology

The condition is rare. It is congenital and inherited as a mendelian recessive, often in the offspring of consanguineous parents.

Pathology

Homogentisic acid is a strong reducing agent which, when oxidized, is converted to a dark pigment. The tissues in which it is deposited, particularly sclerae, ligaments, tendons and the cartilage of the ears, the nose, joints and intervertebral disks, become darkened with pigment. The cartilage loses its elasticity, becomes brittle and has poor resistance to mechanical strain. It cracks easily and is worn away by friction and compression. The exposed bony surfaces undergo sclerosis and formation of marginal exostoses, changes typical of degenerative arthritis. The intervertebral disks, particularly in the lumbar area, degenerate and calcify. The disk spaces narrow, and opposing surfaces of vertebral bodies become irregular and sclerotic. A picture of severe degenerative arthritis of the spine results.

Clinical Picture

The onset occurs in infancy and childhood. The urine blackens on standing, and the diapers become stained. Gradually over the years a slate, brown or black pigment appears first about the concha and the antihelix of the ears and in the sclerae but produces no symptoms. Later, symptoms of degenerative arthritis appear in the large joints and the spine. The entire thoracic and lumbar spine displays rigidity, increased rounding of the thoracic spine

[15] Harrold, A. J.: Alkaptonuric arthritis, J. Bone & Joint Surg. 38:532, 1956.

and flattening of the lumbar spine. Often the patient assumes a posture suggestive of Paget's disease. Perspiration stains the clothing.

Roentgenographic Findings

Films of the spine are characteristic. The disks appears as elliptical, thin, calcified wafers. The apposing vertebral bodies are sclerotic and spurred, and other evidence of degenerative arthritis is seen. The large joints show only degenerative changes.

Laboratory Findings

Homogentisic acid is a strong reducing agent which is oxidized on exposure to air and turns the urine black. Heating or the addition of alkalies hastens the reaction. Copper solutions, as Benedict's or Fehling's, are reduced, and an erroneous diagnosis of diabetes may be made. However, fermentation tests are negative, and the plane of polarized light is not rotated. A diagnostic reaction is the bluish-green coloration produced by the addition of a drop of dilute ferric chloride solution to the urine.

Treatment

There is no known treatment for this condition. Reduction of intake of food containing tyrosine and phenylalanine plus a high dosage







Fig. 38. (*Left*) Thoracic spine showing narrowed, calcified intervertebral disks. (*Right*) Roentgenograms of lumbar spine. The disks are narrowed and calcified. (Harrold A. J.: Alkaptonuric arthritis, J. Bone & Joint Surg. 38B:532)



FIG. 39. Osteoporosis, characterized by diffuse demineralization, loss of transverse striations, thinning of longitudinal striations, and decreased width of the cortices. In this case a fracture has occurred in the supracondylar area of the femur as a result of trivial trauma.

of vitamin C will reduce excretion of homogentisic acid but has no effect on the progress of the disease. The body will produce the aromatic acid from endogenous sources.

OSTEOPOROSIS

Definition. Osteoporosis is a diffuse reduction in bone density which is basically due to failure of formation of the protein matrix in which the calcium is laid down. Histologically, this is apparent by either diminished osteoblastic activity or excessive osteoclastic activity. Osteoblasts are rare and little or no new bone apposition can be seen. The cortices are reduced in thickness, and cancellous trabeculae become thinned as marrow spaces are widened.

Etiology. The cause of lack of bone protein varies, and often several etiologic factors are operative in the same patient. Malnutrition causes osteoporosis because insufficient protein is available. At all times there exists a balance of protein anabolism and catabolism in the body. Osseous anabolism cannot be carried on unless protein materials are readily available. In the presence of adequate nutrition, the cause of osteoporosis must be sought in a disturbance of protein metabolism. This can affect all tissues everywhere, or it may be limited to the skeletal system as in osteogenesis imperfecta, or it may be confined to one or a few bones within a limited area. Certain bones become porotic more rapidly than others. Cancellous bone becomes porotic more rapidly than compact bone. When osteoporosis is generalized, it tends to be most pronounced in the spine and the pelvis, as for example in postmenopausal osteoporosis.

The following are the conditions producing osteoporosis. When decreased skeletal density is due to disturbance of calcium metabolism, the condition, by definition, is termed *osteomalacia*.

DISUSE OR IMMOBILIZATION. The building up of bone exists in balance with the tearing-down process. Greater bone production is a response to greater stresses and strains when maintained within physiologic limits. When these forces are diminished or absent, the breakdown process exceeds the rate of bone build up. An immobilized or paralytic limb is a good example of localized porosis of bone. Osteoporosis is generalized when the entire body is inactive, e.g., the prolonged bedfastness of paraplegics. Excessive removal of calcium from bone means that the mineral must be transported elsewhere by the blood stream. As a result, bedridden patients often exhibit extensive deposits of calcium in soft tissues and formation of renal calculi.

PROTEIN DEFICIENCY. Malnutrition at the present time is rare. On the other hand, gastrointestinal disturbances may impair adequate absorption of protein. Conditions which pro-

duce loss of protein from the body cause a state of protein deficiency. Examples are extensive third-degree burns, nephrosis and chronic draining sinuses.

REFLEX DYSTROPHY. Sudeck first described a peculiar mottled osteoporosis of bony structures about an area of trauma. Fractures, sprains, concussion, etc., may initially display a typical picture of local swelling, intense pain, warmth and the reddish discoloration of vasodilatation. Later, with the onset of vasospasm, the extremity becomes cold, purplish and edematous, and the skin is glossy and atrophic. Hyperhidrosis is evident. Theoretically, the traumatized area reflexly stimulates the sympathetics. The local vasodilatation and sluggish blood flow, perhaps by an increase of local acidity, are associated with spotty foci of bone resorption. Similarly, vasodilatation in other instances, as inflammation, vascular tumors and repair of fractures, produces local osteoporosis.

HORMONAL CAUSES. The formation of protein matrix is dependent upon gonadal hormones. When sex hormones are inadequate, protein anabolism is reduced, while catabolism continues unabated. The condition is more apt to occur in women during the climacteric, especially if the menopause is sudden, as when surgically induced. Less often does it occur in men and then only as a result of total loss of testicular function. Androgen depletion in males is very gradual, inasmuch as the adrenals as well as the testicles produce this steroid hormone. Furthermore, physical activity is greater in men. As a general rule, when osteoporosis is observed in a male, some cause other than hormonal deficiency must be sought.

The sugar or "S" hormone of the adrenal cortex is essential to the process of converting proteins to glycogen. In hyperfunction of the adrenal cortex, whether due to tumor or hyperplasia, an excess of "S" hormone causes excessive breakdown of protein in all tissues, including the skeleton and the musculature. Osteoporosis is, therefore, a feature of Cushing's disease (basophilism of the pituitary) or Cushing's syndrome (primary hyperfunction of the adrenal cortex)

Osteoporosis is part of the hyperthyroid state. Presumably, this develops because an increased catabolism breaks down protein at a faster than normal rate. The result is increased calcium and nitrogen excretion in the urine.



FIG. 40. Senile osteoporosis of spine. Ballooning of disks, biconcave vertebral bodies and pathologic fractures.

SENILITY. With advancing age the rate of new bone formation is reduced, while bone removal continues at a normal pace. Microscopically, osteoblasts are sparse, trabeculae are thin and vertically disposed, and the marrow is fibrofatty and relatively avascular. The condition appears to be a result of many factors, including inactivity, lessened gonadal hormones, dietary lack, etc.

OSTEOPOROSIS OF THE SPINE

The spine is often the site of the most profound changes in generalized osteoporosis and will serve as a typical example of the condition. The characteristic pain and pathologic fractures can affect any bone involved by osteoporosis.

Etiology. The causes include disuse or immobilization, protein deficiency, hormonal causes and senility. The condition most commonly affects women following the menopause.

Pathology. As the vertebrae become soft and fragile they undergo typical changes. The vertebral bodies throughout the thoracic spine as a result of anterior compression forces become wedge-shaped. Those in the lumbar area are exposed to the expansile force of the intervertebral disks so that superior and inferior surfaces of the bodies become indented. The



FIG. 39. Osteoporosis, characterized by diffuse demineralization, loss of transverse striations, thinning of longitudinal striations, and decreased width of the cortices. In this case a fracture has occurred in the supracondylar area of the femur as a result of trivial trauma.

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PROTEIN DEFICIENCY. Malnutrition at the present time is rare. On the other hand, gastrointestinal disturbances may impair adequate absorption of protein. Conditions which pro-

8

Infections of Bone

OSTEOMYELITIS

The term "osteomyelitis," taken literally, implies inflammation of bone and its marrow regardless of whether it is due to pyogenic organisms, tuberculosis, syphilis, a specific virus or the presence of a foreign body such as shrapnel. However, universal acceptance of the term is applied only to infection by pyogenic bacteria, less commonly to the granulomatous inflammation of tuberculosis and syphilis. The infection involves the marrow spaces, the haversian canals and the subperiosteal [illegible]

It is [illegible]

trose [illegible]

tifier [illegible]

resorbed by osteoclasts and actively reconstructed by osteoblasts.

ACUTE OSTEOMYELITIS

Acute osteomyelitis is a rapidly destructive pyogenic infection, usually hematogenous in origin, occurring most frequently in infants [illegible] fatally.

Etiology

A. Predisposing Causes.

1. *Age:* Infancy and childhood, rarely at other ages.

2. *Sex:* Males predominate 4:1.

3. *Trauma:* History of a direct blow is frequently elicited.

4. *Location:* Metaphysis of a long bone. The most actively growing end of the bone, e.g., upper end of tibia, lower end of femur.

5. *Poor nutrition, unhygienic surroundings.*

6. *Antecedent focus* of infection, e.g., boil, tonsillitis.

B. Exciting Causes.

1. *Hemolytic staphylococcus aureus* most common.

2. *Streptococcus* less common.

3. Other organisms rare—*B. coli, B. aerogenes capsulatus* are introduced from septic wounds. Also pneumococcus, typhoid, etc.

Pathogenesis[1]

Although acute osteomyelitis may be initiated by introduction of bacteria from the outside through a wound or continuity from a neighboring soft tissue infection, hematogenous spread from a pre-existing focus is by far the most common route of infection. A septicemia or bacteremia is invariably present. An infective embolus enters the nutrient artery and is trapped in a vessel of small caliber. Most of the small end arteries and capillaries are located in the metaphysis adjacent to the epiphyseal plate. This satisfactorily explains the predilection of the metaphysis to infection. Phemister showed that the predisposition to infection in the metaphysis of a long bone is in direct proportion to the rate of growth at that area and to the size of the bone. The longer and larger the bone, the more susceptible it is to acute osteomyelitis, particularly at its more rapidly growing end. Thus, the upper [illegible]

causes locus minoris resistentiae and provides an excellent culture medium.

The infective embolus, which contains viru-

[1] Luck, J. V.: Bone and Joint Diseases, Springfield, Ill., Thomas, 1950

result is a series of biconcave vertebral bodies and widened disk spaces Albright likens the appearance to that of the spine of a codfish. The margins of the bodies become radiologically *indistinct*, [illegible]
ticularly the tra [illegible]
usual site for a [illegible]
ture is about the dorsolumbar junction. The occurrence of a fracture elsewhere is cause for suspecting other pathology.

Clinical Picture. Patients with osteoporosis complain of pain in the bones, particularly in the back. The individual has a markedly rounded thoracic spine, a stooped habitus and a shortened stature. Deformities result from spontaneous fractures. A compression fracture may result from a trivial trauma, such as opening a window. Frequent acute attacks of backache suggest minimal fractures which cannot be identified in roentgenograms. The acute onset, the point of tenderness, and relief with recumbency are the chief diagnostic signs.

Roentgenologic Findings. Diffuse radiolucency is the main feature. The transverse trabeculations have disappeared, and the remaining vertical ones are thinned. Articular cortices are indistinct. Thoracic vertebral bodies are wedge-shaped, lumbar bodies are biconcave, and lumbar disks are ballooned. Old or recent compression fracture may be observed.

Laboratory Findings. Serum calcium, phosphorus and alkaline phosphatase are normal Urinary excretion of calcium and phosphorus may be slightly greater than the intake. 17-ketosteroid urinary excretion may be increased as a result of adrenal hyperfunction.

Differential Diagnosis. The main conditions to be differentiated are osteomalacia and osteitis fibrosa generalisata which likewise cause generalized demineralization of the spine. The former is characterized by low serum phosphorus and normal calcium, the latter by a low serum phosphorus and increased alkaline phosphatase.

Other causes to be considered are: osteogenesis imperfecta, blood dyscrasia, multiple myeloma, Gaucher's disease, hyperthyroidism and Marie-Strümpell disease.

Treatment. Estrogen and androgen are given for their protein anabolic effect. Premarin (1.25 mg.) is given orally 3 times a day for 4 weeks, 1 week is skipped, and treatment is repeated. Stoppage of estrogen allows withdrawal bleeding and avoids overstimulation of the breast and the uterus. Estrogenic therapy must be continued for a long time, even years. Androgen may be administered as "linguasorbs" (6 mg.) by absorption through the buccal mucous membrane 3 times a day. Instead a repository form of both hormones may be injected at bimonthly intervals (Deladumone). These hormones have a sodium retentive effect; a diet poor in salt and high in protein is necessary. An excess of calcium and vitamin D must be avoided, since these patients do not lack these components. An excess will aggravate the hypercalciuria and lead to renal calculi. Excessive immobilization is avoided to deter atrophy of disuse. Back extensor exercises are valuable even for elderly patients. They provide activity necessary for new bone formation and reduce compression force on the bodies of the vertebrae anteriorly.

Estrogenic therapy decreases calcium and phosphorus excretion in all types of osteoporosis except the idiopathic types. Its effects are manifested within 6 days, are maximal in 30 days and persist for 30 to 50 days after the therapy is stopped. The serum inorganic phosphorus which tends to be high in postmenopausal states falls during estrogenic therapy.

Androgen likewise decreases calcium and phosphorus excretion. It retards *urinary nitrogen excretion*. These effects persist for a long period of time after the androgen administration is stopped.[16]

[16] Reifenstein, E C, Jr Metabolic disorders of bone *in* Harrison, T R (ed), Textbook of Internal Medicine, Sect. 3, pp 651-682, Philadelphia, Blakiston, 1950.

lent organisms in large numbers, blocks a small vessel, and a small area of bone becomes necrotic. An active hyperemia develops in the vicinity, and serum and polymorphonuclear leukocytes are poured out as an exudate to combat the invaders. The hyperemia and enforced immobilization occasioned by pain effects decalcification of the surrounding bone. Proteolytic ferments formed by leukocytes destroy bacteria, necrotic bone and medullary elements. The debris and the exudate increase in amount and effect pressure within the rigid unyielding walls of bone. Other blood vessels are compressed, and further bone necrosis ensues. The exudate follows paths of least resistance, mainly through the haversian and the cortical Volkmann canals to enter the subperiosteal space. Here an accumulation of exu-

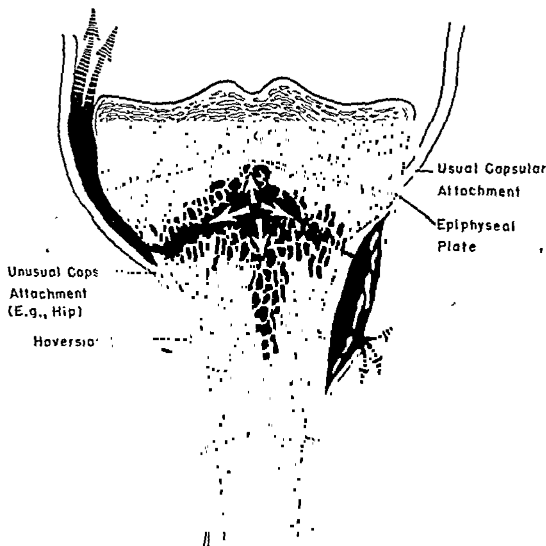


Fig. 41. Spread of exudate in acute hematogenous osteomyelitis. Infection occurs in the metaphysis The cartilaginous epiphyseal growth plate constitutes a barrier to spread toward the epiphysis. The central area of destruction is surrounded by trabeculations rendered atrophic by hyperemia and destruction by proteolytic enzymes The exudate follows the path of least resistance downward along the medullary spaces and laterally through haversian and the Volkmann canals into the subperiosteal space. The attachment of the capsule about the periphery of t[illegible] as in the case of [illegible] metaphysis. Elev[illegible] osteogenesis in an effort to wall off infection The exudate re-enters the bone through haversian channels, or the periosteum is perforated with spread into the surrounding soft tissues.

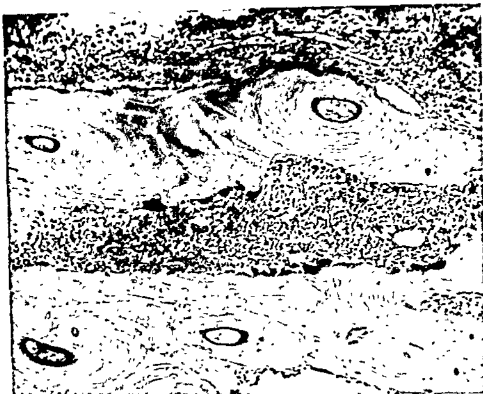


FIG. 43. Chronic osteomyelitis. The marrow spaces are occupied by inflammatory exudate (whose cells are composed chiefly of round cells) and detritus. The bone is dead, as evidenced by empty lacunae, and is being actively resorbed by osteoclasts.

necrosis of bone is inevitable. Upon the mere suspicion of acute bone infection, no harm is done by removing the cortex and providing free access to the outside. A waiting period of 24 to 48 hours is permissible (1) if general supportive treatment by transfusions and fluid is necessary to transform a severely ill child into a better surgical risk, and (2) if symptoms are not too severe and antibiotics alone offer a chance of cure.

Technic. The periosteum is incised longitudinally over the point of maximum tenderness. It is stripped laterally for a half inch on either side. Extensive stripping will jeopardize periosteal blood supply to the cortex. If pus is subperiosteal, the site of bone infection is evidenced by pitting and points of exudation through the cortex. Before the stage of subperiosteal abscess formation, the cortex appears normal. Drill holes are made in various directions until the intraosseous exudate is localized. Then an adequate rectangle of cortex is outlined by multiple drill holes, and removal is completed with the osteotome. Curetting of the spongiosa is avoided for fear of disseminating the infection. The cavity is loosely packed with plain gauze to provide a drain and to keep the wound open. A catheter is introduced into the cavity for local instillation of antibiotics. A cast is applied, and the extremity is placed in a position to provide dependent drainage.

Usually pain is relieved immediately, the temperature falls, and the acutely ill patient is transformed dramatically to a well individual. The wound should be allowed to heal by secondary intention. If destruction is extensive, immobilization by a cast is continued to prevent a pathologic fracture until new bone formation is sufficient.

CHRONIC OSTEOMYELITIS

When the original acute bone infection has subsided, it may persist as a low-grade infection subject to repeated recrudescences of the acute process over many months or years. Hematogenous infection with an organism of low virulence may be chronic from the onset. Infection introduced through an external wound usually causes a chronic osteomyelitis.

PATHOLOGY[2]

In any infection of bone, there is an attempt at repair which, if incomplete, results in chronic persistence of infection. This repair is accomplished by hyperemia of the surrounding tissue which effects decalcification of the

[2] Hodges, P. C., Phemister, D. B., Brunschwig, A.: Diseases of Bones and Joints, New York, Nelson, 1938.





Fig. 44. Chronic osteomyelitis of the upper femur.

bone Granulation tissue forms and carries in osteoclasts and osteoblasts Necrotic cancellous bone is readily absorbed and replaced by new bone. Dead cortex is gradually absorbed about its surface and is gradually detached from living bone to form a *sequestrum*. The erosive process is brought about by osteoclasis and the action of proteolytic enzymes and causes a jagged irregular surface appearance. Because of loss of blood supply, the sequestrum is not decalcified and appears more dense than the surrounding living bone. Separation of dead from living bone may require several months. Rarely, when infection has been completely eradicated, the necrotic bone does not separate and instead is gradually replaced over many months by the process of creeping substitution. After complete sequestration so that the dead fragment lies free within a cavity, it is less readily attacked by granulation tissue and is absorbed more slowly. This is especially the case when infective exudate fills the cavity. The surrounding living bone attempts to wall off the infection by forming a thick, dense wall, the *involucrum*. When neighboring tissue is destroyed, the surface of the sequestrum remains uneroded. Externally, the periosteum lays down new bone to form an involucrum Rarely, the entire shaft will sequestrate and be enveloped by a new periosteal bone casing which gradually increases in density and thickness. An involucrum usually has multiple openings, the *cloacae*, through which exudate, bone debris and sequestrae find exit and pass through sinus tracts to the surface. Constant destruction of neighboring soft tissues leads to cicatrix formation, and the skin is thin, distorted and easily traumatized, the skin epithelium growing inward to line the sinus tracts. Persistence of drainage irritation may cause an epithelioma of a sinus tract.

After a sequestrum has been discharged or removed, the sinus usually closes, and the cavity may fill with new bone. This is more likely in children. In adults, the cavity may persist and harbor organisms which may reactivate the infection at any time.

In chronic osteomyelitis of long standing, multiple cavities and sequestra exist throughout the bone. The shaft becomes thickened, irregular and deformed.

date gradually strips the periosteum as a subperiosteal abscess. The periosteum in turn attempts to wall off the infection by forming new trabeculae of bone. The underlying cortex, deprived of its periosteal blood supply, becomes necrotic. Perforation of the periosteum permits spread of the infection in the soft tissues along fascial planes.

The exudate may spread down the medullary canal, destroying marrow elements and the blood supply to the cancellous bone and inner aspect of the cortex. In an advanced stage, the cortex may be surrounded externally and internally by pus and deprived of its blood supply, the entire diaphysis sequestrating. It is possible for a small accumulation of pus, by compromising the nutrient artery, to endanger large areas of the diaphysis. The importance of early decompression and adequate drainage is readily apparent.

Dead bone is absorbed by granulation tissue and osteoclastic activity about its surface. When the dead bone is large, it is gradually separated from living bone and is slowly destroyed. Because it has no blood supply, the sequestrum appears to be dense in comparison with surrounding decalcified bone. Spongy bone is absorbed rapidly.

Sp[illegible] joint [illegible] plat[illegible] trate the periosteum, spread along the soft tissues and penetrate the capsule in order to cause a suppurative arthritis. The hip is an exception. The metaphysis lies within the confines of the capsule, and the joint is directly involved.

Healing may take place at any stage due to natural resistance or under the influence of antibiotics. At the earliest stage of minimal destruction, the exudate is resorbed, and new bony trabeculae are formed. Organisms of lesser virulence and inadequate resistance may result in the formation of a persisting abscess which is surrounded by a fibrous membrane and walled off by a ring of dense bone. This is known as a *Brodie's abscess*. The infection may be reactivated at any time. Or the organisms are destroyed, and a cavity containing sterile pus, serous fluid, or fibrous tissue will remain indefinitely.

Necrotic bone becomes absorbed slowly over its surface, is surrounded by granulation tissue, separates from surrounding living bone, and persists until slowly resorbed or extruded. Until this happens, exudate is formed continually and may drain externally. Walled-off areas of infection may undergo recrudescence of activity at any time in later years. This is the stage of chronic osteomyelitis.



Fig. 42. Subacute osteomyelitis. Reactive periosteal new bone completely envelops the shaft. The destructive lesion lies anteriorly.

Clinical Picture

An antecedent infection is usually present. With the onset of osteomyelitis, symptoms of a severely acute illness appear. The child is irritable and restless and complains of headache. Vomiting, convulsions and chills occur. Fever is high, the pulse is rapid, and leukocytosis is as high as 30,000. The extremity is held in semiflexion, surrounding muscles are in spasm, and passive movement is resisted because of pain. Pain quickly becomes excruciating and occasions fits of screaming and crying. At first no swelling is evident, but within a few days the soft tissues about the affected site become edematous and red, indicating subperiosteal abscess formation. Before

periosteal involvement, a localized point of tenderness is found over the affected metaphysis. After inflammatory signs appear, tenderness is quite pronounced. Fluctuation is not elicited until pus has escaped outside the periosteum. An increased effusion in the adjacent joint proves in most cases to be a sympathetic synovitis with sterile clear fluid. If infection and septicemia proceed unabated, the child may become apathetic and unconscious and continue to a fatal termination.

Laboratory Findings

1. **Roentgenograms.** Films are negative within the first week or 10 days. Thereafter a loc [illegible]
in [illegible]
of [illegible]
weeks, the periosteal shadow is elevated at the same level, and multiple laminations of bone deposition parallel with the shaft are seen. Eventually, more spongy trabeculae are destroyed, giving a moth-eaten appearance which extends for a varying distance in the medulla toward the diaphysis. The external and/or internal surface of the cortex may display multiple scalloped erosions. If a segment of necrotic bone is present, it retains its original architecture and appears denser than the surrounding decalcified bone When healing takes place in the earliest stage, the bony architecture is quickly restored.

2. **Aspiration** of subperiosteal pus reveals by culture the infecting organism and its sensitivity to antibiotics.

3. **Blood culture** demonstrates the presence of bacteremia.

4. **Blood count** reveals a polymorphonuclear leukocytosis.

Complications

Spread to surrounding soft tissues may cause suppurative tenosynovitis, suppurative arthritis and thrombophlebitis.

Differential Diagnosis

The main conditions to be ruled out are.

1. **Rheumatic Fever.** The onset is more gradual, and general constitutional symptoms are less acute. Pain and tenderness are less intense and are confined to the joint. Involvement is polyarticular. Response to salicylates and ACTH is dramatic. Antibiotics have no effect.

2. **Ewing's Tumor.** This, too, causes fever, leukocytosis and subperiosteal "onion-peel" bone deposition. However, destruction usually is confined to the diaphysis, is more diffuse, and the tumor responds rapidly to x-ray irradiation. Constitutional symptoms are less intense. Biopsy demonstrates the presence of tumor cells.

3. **Acute Suppurative Arthritis.** Fluid accumulation in the joint occurs earlier, pain and tenderness are definitely limited to the joint, joint movement is greatly restricted, muscle spasm is intense, and aspiration reveals the presence of purulent synovial fluid.

Prognosis

The 20 per cent mortality of past years has been greatly reduced to the point of rarity with the introduction of antibiotics. Rapid cure of infection in the early stage before bone destruction is evident apparently occurs frequently. It is almost impossible to determine the presence of the disease at this stage. It has become uncommon to observe a case after severe bone destruction has occurred. Small areas of necrotic and sequestrated bone may be resorbed and normal bony architecture restored, especially in children. However, a large sequestrum surrounded by a wall of dense bone may retain infective material within its cavity and cause exacerbations of infection and draining sinuses over many years in spite of treatment with antibiotics. Chronic osteomyelitis continues until the sequestrum is resorbed or extruded.

Treatment

Even before the diagnosis can be definitely established, an antibiotic is given immediately in large dosage and continued indefinitely. Penicillin and chlortetracycline are preferred until the organism can be obtained, cultured and tested for sensitivity. Like in any infection in a closed space, immediate provision for drainage is of paramount importance. This must be done at the earliest possible opportunity, even before signs of subperiosteal infection are evident. To wait is to invite disaster Pressure exerted by pus enclosed within a rigid compartment is tremendous, the circulation to the bone is jeopardized, and extensive

Fig. 45. Localized osteomyelitis of the tibia with sequestrum formation.

Clinical Picture

During the period of inactivity, no symptoms are present. The bone is misshapen, and the skin is dusky, thin, scarred and poorly nourished. A break in the skin causes an ulceration which is slow to heal. Muscles are scarred and cause contractures of adjacent joints. A lighting up of infection is manifest by aching pain which is worse at night. The overlying soft tissues become swollen, edematous, warm, reddened and tender. The temperature may be elevated a degree or two. As the infection progresses, a sinus may open and drain indefinitely, extruding small sequestrae at intervals Spontaneous closure of the sinus and subsidence of infection often occur following expulsion of a large fragment.

These recurrent acute flare-ups occur at indefinite intervals over months and years. A sinus may drain continuously. On the other hand, an interval of many years between flare-ups is not unusual. Relapse is often the

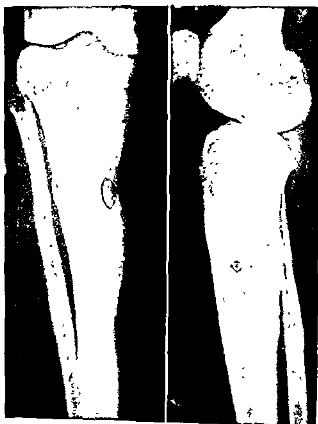


Fig 46. Chronic osteomyelitis. A large sequestrum is enclosed within the cavity.

periosteal involvement, a localized point of tenderness is found over the affected metaphysis. After inflammatory signs appear, tenderness is quite pronounced. Fluctuation is not elicited until pus has escaped outside the periosteum. An increased effusion in the adjacent joint proves in most cases to be a sympathetic synovitis with sterile clear fluid. If infection and septicemia proceed unabated, the child may become apathetic and unconscious and continue to a fatal termination.

Laboratory Findings

1. **Roentgenograms.** Films are negative within the first week or 10 days. Thereafter a localized area of bone destruction is observed in the metaphysis surrounded by a wide zone of decalcified bone. Later, within the next few weeks, the periosteal shadow is elevated at the same level, and multiple laminations of bone deposition parallel with the shaft are seen. Eventually, more spongy trabeculae are destroyed, giving a moth-eaten appearance which extends for a varying distance in the medulla toward the diaphysis. The external and/or internal surface of the cortex may display multiple scalloped erosions. If a segment of necrotic bone is present, it retains its original architecture and appears denser than the surrounding decalcified bone. When healing takes place in the earliest stage, the bony architecture is quickly restored.

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Prognosis

The 20 per cent mortality of past years has been greatly reduced to the point of rarity with the introduction of antibiotics. Rapid cure of infection in the early stage before bone destruction is evident apparently occurs frequently. It is almost impossible to determine the presence of the disease at this stage. It has become uncommon to observe a case after severe bone destruction has occurred. Small areas of necrotic and sequestrated bone may be resorbed and normal bony architecture restored, especially in children. However, a large sequestrum surrounded by a wall of dense bone may retain infective material within its cavity and cause exacerbations of infection and draining sinuses over many years in spite of treatment with antibiotics. Chronic osteomyelitis continues until the sequestrum is resorbed or extruded.

Treatment

Even before the diagnosis can be definitely established, an antibiotic is given immediately in large dosage and continued indefinitely. Penicillin and chlortetracycline are preferred until the organism can be obtained, cultured and tested for sensitivity. Like in any infection in a closed space, immediate provision for drainage is of paramount importance. This must be done at the earliest possible opportunity, even before signs of subperiosteal infection are evident. To wait is to invite disaster. Pressure exerted by pus enclosed within a rigid compartment is tremendous, the circulation to the bone is jeopardized, and extensive

until the defect is filled with granulation tissue. Next, a split-thickness skin graft (about 0.012 inch thick) is placed over the granulations, and a pressure dressing is applied. After the take, the area is observed over a period of months to ascertain whether or not the infection is quiescent. Finally, if the bony defect is [illegible] a bone-grafting procedure. It is preferable to create a pedicle flap and transfer it gradually until it becomes very viable and well-nourished before using it to replace the split-skin covering. After several months, the thick graft will have good circulation and will heal readily following the bone-grafting procedure. Finally, the bone defect is approached, if possible, through normal skin. All scar tissue is excised, and the walls of the cavity are curetted until cancellous, bleeding bone is exposed. Multiple [illegible] is applied.

SCLEROSING OSTEOMYELITIS OF GARRÉ

(Idiopathic Cortical Sclerosis)

This disease consists of the gradual development of a spindle-shaped sclerotic thickening of the cortex of a long bone usually confined to one side of the shaft. Infants and children are affected most frequently, and the tibia is the usual site. Pain is constant, dull, boring, and worse at night. A history of trauma is often elicited. Clinically, one finds a diffuse bony enlargement which is only slightly tender. The overlying soft tissues appear normal or occasionally may be slightly warm and reddened. Constitutional symptoms are absent, and the blood picture is normal.

The roentgenologic picture shows a gradual thickening and increased density of the cortex. Within the sclerotic area, a minute focus of decreased density may be observed, particularly when films are taken in various planes and with varying intensities of exposure.

Microscopic examination in most cases fails to reveal evidence of inflammation. The occasional finding of an osteoid osteoma type of lesion suggests this as the causative lesion. Failure to remove the nidus may explain persistence of symptoms. Before excising a section of cortical bone, an attempt should be made to identify and localize a nidus. If symptoms persist, roentgenographic study is repeated, and further bone resection is attempted. Antibiotics are valueless.

The term "osteomyelitis" should be discarded. In the absence of a discoverable etiologic lesion, the general all-inclusive label "idiopathic cortical sclerosis" is suggested.



FIG. 47. Synovial tuberculosis.

TUBERCULOSIS OF BONES AND JOINTS

Tuberculosis is a chronic infectious disease caused by the tubercle bacillus. Involvement of bones and joints is secondary to lesions elsewhere. Therefore, reduction in the incidence of tuberculosis by health measures and lessening of the severity of the disease, particularly by antibiotics and chemotherapeutic agents, has made tuberculosis of bones and joints an uncommon condition.

result of poor bodily condition and lowered resistance. Recurrent toxemia over a long period of time will eventually cause debilitating and sometimes fatal amyloidosis.

Complications of chronic osteomyelitis include a reduced rate of growth, pathologic fracture, bone lengthening, muscle contracture, epithelioma and amyloidosis.

The causative organisms are usually the staphylococcus and the streptococcus. Contaminating saprophytes are invariably present and produce penicillinase, which counteracts the effectiveness of penicillin. These bacteria include Clostridia, Aerogenes, Proteus, Pyocyaneus and *Escherichia coli*.

Roentgenographic Findings

In the early stages, the bone appears motheaten and osteoporotic, and areas of sclerosis develop. The periosteum is elevated by subperiosteal laminations of new bone, which become progressively thicker and dense. Sharply delineated areas of density, the necrotic bone, are accentuated by surrounding decalcification. Gradually, each necrotic dense area becomes surrounded by a white ring representing reactive new bone formation, the involucrum. There then develops a narrow zone of diminished density between necrotic and living bone, signifying absorption about the surface of the sequestrum and separation from living bone. If this narrow zone of bone resorption does not appear, infection is probably not present, and the central density represents an area of aseptic necrosis which will be replaced by creeping substitution. Many areas of increased and decreased density may occur throughout the bone, the shaft becoming enlarged and misshapen. A sequestrum may not be visible because of overlying dense bone unless x-ray exposures are made with varying intensities and from various projections.

Treatment

Defense mechanisms of bone constantly strive to absorb or extrude the sequestrum. Exudate is constantly formed under pressure, which further compromises the circulation and spreads the infection. In addition, the hard sclerotic wall, which likewise is infected, prevents obliteration of the cavity in which organisms can reside indefinitely. The wall acts as a barrier, preventing access of antibiotics to the cavity. It is obvious that cure of infection requires removal of the sequestrum and excision of infected granulation tissue, scar and thick involucrum. By providing a residual bed of normal, bleeding, cancellous bone, bone regeneration and healing is rapid. Removal of bone should not be extensive for fear of inducing a pathologic fracture. If the parent bone is involved diffusely and severely, a conservative approach is best. Only the offending sequestrum is removed. When infection is extensive and uncontrollable, amyloid disease is a definite danger. Amputation is best. The following technic may be applied to chronic osteomyelitis regardless of whether it is caused by penetrating wounds, compound fractures, or following an acute bone infection.

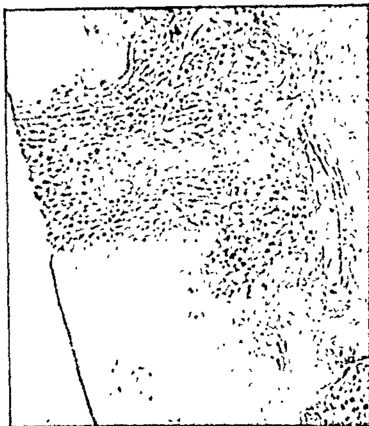
Technic.[3, 4] Preoperatively, the patient's condition is improved by multiple blood transfusions and a high protein diet. The exudate is cultured, and the organisms are tested for sensitivity to various antibiotics. The offending organism is usually the staphylococcus, but saprophytes and secondary invaders are also present. These are of importance, as they produce penicillinase which destroys the antibiotic and allows the staphylococcus to survive. These contaminants most commonly include *Escherichia coli* and *Pseudomonas aeruginosa*. Often these can be eliminated by local application of streptomycin. Most strains of *Ps. aeruginosa* are sensitive to aerosporin. Otherwise, the main infecting bacteria is combated by the indicated antibiotic.

A sufficient section of cortex is removed to permit free drainage, and the edges are saucerized. All sequestra, scar tissue and surrounding dense bone is excised until a bed of raw, bleeding cancellous bone remains. Any tissue causing the mere suspicion of infection or creating doubt as to viability should be removed. The [illegible] n [illegible] e [illegible] ays

[3] Speed, J. S., and Smith, H.: Campbell's Operative Orthopedics, ed. 2, p. 1147, St. Louis, Mosby, 1949.

[4] Hazlett, J. W.: Treatment of osteomyelitic defect by cancellous bone grafts, J. Bone & Joint Surg. 36B: 584, 1954.

PLATE 10. Tuberculosis. The tuberculous granulation tissue is seen undermining and separating the articular cartilage from the bone. It does not directly destroy cartilage but instead enters the joint through a split in the cartilage. Several typical Langhans giant cells are present. (× 210)



necrosis. The adult infection is a fresh one which varies in its ability to overcome the immunity produced by the first lesion.

The Tubercle. The initial response, especially in reinfection (allergic inflammation of an already tuberculous animal), is polymorphonuclears which are rapidly replaced by mononuclears (macrophages and monocytes), which are highly phagocytic members of the reticuloendothelial system After phagocytosing the bacilli, the latter break down, and the lipid is dispersed throughout the cytoplasm, the mononuclear being transformed into the *epithelioid cell.* This cell, characteristic of the tuberculous reaction, is a large pale cell with a large vesicular nucleus, abundant cytoplasm, indistinct margins, and processes which seem to pass between the cells, forming an epithelioid reticulum. The characteristic Langhans giant cell with its peripherally placed nuclei are probably formed by fusion of a number of epithelioid cells. They are not formed until caseation necrosis has occurred, often they contain tubercle bacilli. Their function is to digest and remove dead tissue. They occur in other chronic inflammations (syphilis, actinomycosis). After a week lymphocytes appear and form a ring about the periphery of the lesion. These are one of the sources of gamma globulin, the immune bodies. This mass of newly formed cells constitutes the translucent nodule known as the tubercle. Several small tubercles may fuse to form a larger one. *Caseation necrosis,* which is a coagulation necrosis formed by liberation of the protein fraction of bacilli, begins at the second week. The homogeneous center stains red with eosin, surrounded by pale epithelioid cells with one or more giant cells and ringed by a zone of dark-blue lymphocytes. The caseous material softens and liquefies.

The future course of the tubercle varies: (1) It may resolve completely. (2) Fibrous tissue may encircle the lesion, and lime salts may be deposited in the central caseous substance. (3) A low-grade inflammation characterized by many tubercles without proceeding to caseation and encircled by granulation tissue, may develop. This fibrosing hyperplastic form is most common in synovial membrane. (4) Infection may spread throughout the tissue with formation of many more tubercles. (5) There may develop an acute inflammatory

Etiology

Exciting Factor is the tubercle bacillus.

Predisposing Factors:

Constitutional: inadequate diet, fatigue, poor sanitation.

Race: dark-skinned races are predisposed, i.e. Negroes, Mexicans, Orientals and American Indians.

Trauma: direct violence to a bone or a joint preceding infection suggests formation of a *locus minoris resistentiae.*

Age: infants under 2 years of age do not withstand infection well. Infection between 2 and 15 years is usually relatively benign. Beyond 15 the disease is severe and may be fatal.

Disease: measles and chickenpox can provoke infection.

Puberty and *pregnancy* can reactivate tuberculosis.



Plate 8. Tuberculous destruction of an interphalangeal joint.

Pathology[5]

Infection in bone and synovial tissue invokes the same response as in the lung except for variations due to the character of the tissue. The initial infection occurs in the lung (human type) or the intestine (bovine type), usually in children under 2 years of age. A natural immunity develops and heals the tubercle at the site of invasion and regional lymph nodes (primary complex). However, allergy to future infection has developed. Therefore, at the *second infection,* which may occur years later, the *allergic response* is an acute inflammatory one, prompt in appearance and marked by outpouring of polymorphonuclear leukocytes and plasma and by massive

[5] Boyd, W Textbook of Pathology, Philadelphia, Lea & Febiger, 1953.

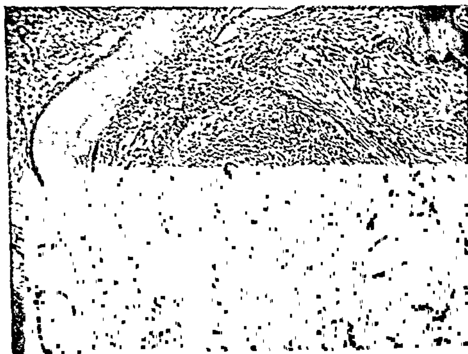


Plate 9. Typical tubercle, showing epithelioid cells, an encircling ring of round cells and fibrous tissue, and the characteristic Langhans giant cell. (× 90)



FIG. 48. Tuberculosis at the end of a long bone and involving the joint. Between the thin trabeculae, some of which are necrotic, are seen tubercles, granulation tissue and cellular infiltrate, all of which extend to but do not directly invade the articular cartilage. Similarly, the heavily cellular tuberculous tissue overlying the articular cartilage does not seem to affect the latter. (× 35)

When the lesion in bone is chiefly exudative, the allergic reaction is present, the acute hyperemia causes decalcification in a localized area, revealed in roentgenograms as an osteolytic lesion. If healing occurs rapidly with absorption of exudate, recalcification takes place with restoration of bony architecture.

When caseation is present, the process is less acute, the coagulation necrosis and peripheral fibrosis imply a reaction of immunity. The central osteolytic area seen in roentgenograms is replaced by an increased density as healing and calcification are taking place. During the early necrosing stage, the surrounding bone undergoes hyperemia and decalcification, but not as severe as in the central area. Occasionally, with healing the perifocal bone may undergo hypertrophy similar to the involucrum in chronic osteomyelitis.

Abscesses in the soft tissues have a tendency to migrate along fascial planes and erupt at a distance from the original focus. This is seen best in the spine where the infective material may enter the fascia enveloping the psoas muscle and erupt at the groin.

The areas of predilection occur in the following order of frequency: spine, hip, knee, ankle, tarsus, shoulder and elbow.

Clinical Picture

Tuberculosis of a bone or a joint is a low-grade slowly progressive infection. The degree

reaction, when infection is virulent or massive, an exudative outpouring of polymorphs and serofibrinous fluid. This is seen most commonly in reinfections.

Skeletal and Articular Involvement. This occurs during a second infection, the period of allergy. Dissemination takes place through the blood stream and affects almost all the bones, the immunity apparently healing most of the lesions. On the other hand, the allergic exudative response may be extreme, overcoming the immunity, and a fatal miliary tuberculosis may ensue. Most commonly, the defense is sufficient to overcome all but a few sites. Implantations occur both in bone and synovial membrane. If only bone or synovium is infected alone, the one generally infects the other in a short time. Typically, an active focus is set up in a metaphysis (in a child) or in an epiphysis (in an adult) where the acute exudative reaction may cause local necrosis until caseation occurs. The intense hyperemia causes marked decalcification locally, a characteristic well seen in roentgenograms. Granulation tissue about the site constitutes the fibroblastic healing response. It brings in tremendous numbers of mononuclears and attempts to delimit the process by fibrosis. If unsuccessful, the destruction extends up and down the shaft and peripherally to reach the subperiosteal space. The periosteum may react to a superficial cortical lesion by producing new periosteal bone. The exudate may penetrate outward through the soft tissue to exit through the skin as a fistula chronically draining caseous material, particles of bone, and partially liquefied, thick, grayish yellow substance. Before this abscess erupts, it distends the skin, which appears thinned, slightly darkened, but without acute inflammation (cold abscess). Secondary infection of the sinus tract is frequently superimposed The prolonged drainage causes amyloidosis and death.

Cartilage is resistant to tuberculous destruction. Therefore, the epiphyseal plate is not destroyed. However, the granulation tissue may invade the area of calcified cartilage and interfere with longitudinal growth. Asymmetrical impairment of this growth process leads to deformity. Metaphyseal infection reaches the joint through the subperiosteal space, from which it penetrates the capsular attachment about the joint. In the adult, after complete ossification of the epiphysis, the infection spreads in the subchondral area to the periphery where the synovium joins the cartilage; then it enters the joint. Destruction of subchondral bone loosens the attachments of the articular cartilage, which may become displaced into the joint cavity.

The synovium may be originally infected and the bone secondarily infected by a reverse process of the one just described. The most common type of synovial infection is one which is low grade. The membrane is swollen and congested with granulation tissue. The articular surface is studded with many translucent tubercles, and deposits of fibrin cover many areas. The joint fluid usually is moderately increased and clear. It contains rice bodies, which are small accumulations of fibrin, and pieces of articular cartilage. Caseation necrosis of the synovium and the capsule is rare. The hydrarthrosis may also occur without the synovium's actually being infected. In this case the inflammatory synovitis and the increased joint fluid is a nonspecific reaction to an adjacent infection in the neighboring bone. Therefore, *a negative synovial biopsy does not necessarily rule out tuberculosis.* Tuberculosis of the synovium generally heals by fibrosis and marked thickening of the membrane The granulation tissue extends from the synovium over the articular cartilage at the periphery where as a pannus it seems to erode the cartilage. When a destructive caseating lesion of bone penetrates the joint, destruction seems to involve the synovium too rather than the usual low-grade fibrosing process. A chronically draining sinus with secondary infection of the joint and generalized amyloidosis may constitute a terminal condition. If the defense reaction overcomes the joint infection, granulation tissue bridges the joint cavity, and a fibrous ankylosis results.

A focus of caseation necrosis may be walled off effectively and a clinical cure achieved. However, viable tubercle bacilli remain indefinitely—a potential source of exacerbated activity and reinfection at a later date. As in other types of necrosis, calcium salts frequently are deposited in the caseous material. In roentgenograms this gives the appearance of a sequestrum.

its value lies in a negative reaction, ruling out the disease. A positive reaction in a known previously negative individual is diagnostic.

Guinea Pig Inoculation. Guinea pigs have no natural immunity to tuberculosis and rapidly succumb. The pus is injected intraperitoneally, and examination discloses tubercles 5 to 8 weeks later.

Culture. This requires from 5 to 30 days. It should be remembered that the serous exudate of a joint may be a nonspecific inflammatory response to neighboring infection and therefore may not contain bacilli.

Biopsy. Microscopic tissue examination reveals typical tubercles. A regional lymph node should be removed for examination.

Blood. A leukocytosis may be present, particularly in the more intense inflammatory types with constitutional symptoms. A relative lymphocytosis is often found.

Sedimentation Rate. This is more likely to be normal in mild granular types of infection. The acutely inflammatory exudative infection gives the highest values.

Roentgenographic Findings

Osteoporosis is the first sign of active infection. It may also occur as a nonspecific response to an adjacent infection of the joint. The reactive hyperemia which is most intense in exudative ir [illegible]
which may be [illegible]
osteolysis. The [illegible]
is lost. When one tarsal or carpal bone is infected, the remaining tarsals and carpals rapidly decalcify [illegible]

and metaphyseal zones. Its rapid spread indicates an intense exudative reaction. Conversely, recalcification means reduction of activity of the disease.

Swelling of the synovial shadow indicates a hydrarthrosis. This results from (1) primary synovial infection or (2) a nonspecific inflammatory reaction to a neighboring bony focus which may not yet be visible in roentgenograms.

Small zones of clearly defined subnormal density in bone indicates granular foci. These osteolytic areas are surrounded by diffuse osteoporosis. As caseation takes place the osteolytic focus is very evident. When healing comes into play, the perifocal bone becomes thickened as a heavy overcalcified ring. If the central decalcified area is merely exudative, healing results in reossification and once again the trabeculae are apparent. More commonly, a central caseation exists in which calcium is deposited and gives the dense image of a sequestrum. This is surrounded by an osteolytic ring representing the fibrous wall, and beyond this the bone is osteoporotic, normal, or dense, depending on the defense reaction. The final picture may resemble osteomyelitis.

In destructive arthritis, necrosis and separation of articular cartilage cause joint space narrowin [illegible]
ragged [illegible]
densities [illegible]
abscess [illegible]
paravertebral shadow about the spine in the thoracic area and widening of the psoas shadow in the lumbar area. Sinuses are traced by injection of radiopaque substance.

[illegible]

growth plate may stimulate longitudinal growth. If it encroaches on the area of endochondral ossification, growth is irregularly retarded, and deformity results. When the focus [illegible]
seen. [illegible]
bones [illegible]
bone may encircle and enlarge the entire diaphysis.

Course and Prognosis

The newer forms of antibiotics and chemotherapeutic agents have greatly improved the outlook in tuberculosis of bones and joints. In the mild granular infections, healing can often be accomplished without residual joint scarring and ankylosis. These substances permit aggressive resection of larger caseous destructive foci from which later infections may originate. The fatal types of meningitis and miliary tuberculosis are now rare.

The tendency is toward healing by fibrosis Mild granular types heal slowly. Exudative caseous lesions heal less readily. An epiphyseal caseous lesion may spread into the joint

of local and general reaction depends on the intensity of infection and the defensive response.

Characteristics of the disease:

1. *An insidious onset*

2. *Monoarticular or mono-osseous* involvement

3. *Other visceral lesions,* e.g., pulmonary, intestinal, renal

4. *Other family members may have tuberculosis*

5. *Trauma* to involved region often precedes onset

6. *Local symptoms and findings:* doughy swelling (caused by synovial inflammatory swelling), slight pain and tenderness, muscle spasm, night cries in children (due to relaxation of muscle spasm which allows painful motion), stiffness early, later actual limitation of active and passive motion due to fibrous ankylosis, slight warmth about involved site, increased joint fluid (due to tuberculous synovitis, or nonspecific reaction to neighboring bone infection), limp, muscle atrophy which may be quite marked and occurs early.

7. *Constitutional symptoms:* low-grade fever, especially in the afternoon, anorexia, weight loss, night sweats, tachycardia, anemia

Types of the Disease. For purposes of description, two clinical types are identified: (1) the granular, mild, nondestructive, fibrosing; and (2) the caseous and exudative, destructive, abscess-forming. Actually, both types occur together, one predominating over the other.

GRANULAR OSSEOUS TYPE. Bone involvement, usually at the metaphysis or the epiphysis, often follows trauma The onset is insidious with limp, vague mild pains and fatigue. Hydrarthrosis of the adjacent joint is nonspecific and appears late in the day after activity. The periosteum may be palpably thickened, and the overlying soft parts slightly warm and tender. Joint movement may be restricted because of hydrarthrosis Muscle atrophy rapidly appears. Constitutional symptoms are rare. Infrequently, a fluctuant swelling indicates abscess formation.

CASEOUS AND EXUDATIVE OSSEOUS FORM Here the onset is less insidious and is associated with marked constitutional symptoms as fever, night sweats, weight loss and anorexia. Pain is more intense and particularly severe at night. Muscle spasm is marked. The overlying soft tissues are warm, swollen, indurated and quite tender. The tendency to abscess formation is greater. Its appearance is preceded by redness, increased heat, and distention of skin which becomes thin and shiny. Rupture of the abscess produces a fistula which drains pus, caseous material and sequestrae for many months. Secondary infection perpetuates the drainage. When the caseous material penetrates the joint, a severely destructive arthritis ensues.

GRANULAR SYNOVITIS. This is the usual type when the joint is predominantly involved, particularly in children. Frequently recurring mild hydrarthroses with little or no pain characterize the insidious onset. Constitutional symptoms are mild. Recurrences become more frequent and persistent. Muscle atrophy gradually appears. In time the joint fluid lessens, and the synovial membrane thickens. Motion becomes limited, particularly at the extremes. The synovitis may last for years without involving the bone. Rarely, it may be converted into the caseous form with increase of local and constitutional symptoms. Eventually, contracture and subluxation occurs.

EXUDATIVE SYNOVITIS. The onset may be acute with intense inflammatory signs and general symptoms. Fever rises up to 101°, and the general condition is poor. Movements are very painful. The regional lymph glands are swollen. The soft tissues about the joint are diffusely swollen and very tender. The condition changes rapidly to exudative caseous involvement of soft tissues and bone, the acuteness subsides, and the tendency is to ankylosis. An abscess usually ruptures externally, and necrotic cartilage is lost. The draining fistula invites secondary infection which over the years leads to amyloidosis.

LABORATORY DIAGNOSIS

The following methods are used most commonly:

Tuberculin Reaction. This depends on an allergic inflammatory response to an antigen. The intradermal method (Mantoux) is best. A positive reaction indicates only that tuberculosis has been present in the past. Therefore,



Fig. 49. Congenital syphilis. Moth-eaten appearance of the metaphyses whose epiphyseal borders are dense and sawtoothed. Periosteal ossification of shafts.

involves the epiphyseal areas. The infection may then extend into the adjacent joint and cause destruction of cartilage and other soft tissues. Sinus formation is followed by secondary purulent joint infection. Healing takes place by fibrosis, but bony ankylosis does not occur.

Early Congenital Syphilis (Parrot's Pseudoparalysis)

The medulla is always infected in the syphilitic newborn Because capillary blood supply is richest in the metaphysis adjacent to the epiphyseal plate, this is the site of maximal involvement. Endochondral ossification is interfered with. The infection, round cells, and granulation tissue invade the zone of calcified cartilage which therefore fails to resorb and accumulates in large amounts. The infected granulation tissue interferes with the laying down of osteoid tissue so that the trabeculae when formed are sparse and irregular. The proliferating cartilage piles up at the epiphyseal plate. These events explain the roentgenographic appearance of a widened translucency of cartilaginous epiphysis and plate, a widened zone of increased density (calcified cartilage) and an irregular sawtoothed contour of the proximal border of the metaphysis, and osteoporosis just beyond the dense zone. The junction between the epiphyseal plate and calcified cartilage is fragile, and fracture with epiphyseal separation often occurs.

Mild metaphyseal infection may stimulate endochondral ossification so that the extremity often displays rapid growth. On the other hand, virulent destructive infection may destroy the growth plate sufficiently to interfere with growth.

termined by the *lymphocyte-monocyte ratio*.[11] When this is greater than 5, a state of good resistance is indicated. On the other hand, a low lymphocyte and high monocyte count indicate a dangerous state of hypergy which favors development of serious complications.

A trial of conservative treatment is justified in cases of low-grade infection. Recalcification and restoration of bone architecture indicates that healing is taking place. When a bone contains an obviously encircled area of caseation, particularly when abscess formation threatens the integrity of neighboring structures, and when continued drainage is progressively debilitating the patient, removal of the infected focus is indicated. The following procedures are employed:

1. *Excision of the focus*
2. *Excision of the entire bone*, e.g., a tarsal bone
3. *Arthrodesis* to put the part at rest.
4. *Drainage and curettage of the abscess*
5. *Amputation* when destruction is extensive

SYPHILIS OF BONES AND JOINTS

The bone, the bone marrow and the periosteum are favored sites of involvement in syphilis. However, with the advent of antibiotics and supervised prenatal care, luetic lesions have become rarities The spirochete *Treponema pallidum* is highly susceptible to penicillin, so that syphilis of the newborn and tertiary lesions of the adult are becoming extinct However, an occasional condition requires identification and differentiation from other diseases.

Pathology

The basic pathology in the skeleton is similar to that of other tissues The spirochete is blood-borne and lodges within the medulla at the site of vessels of smaller caliber, particularly at the metaphysis. Its presence calls forth a low-grade inflammatory response consisting of vasodilatation and a local outpouring of serum and mononuclear cells (lymphocytes and plasma cells). The cells characteristically congregate about blood vessels. If defenses are adequate, the bacteria may be destroyed and the exudate resorbed, the tissues returning to normal; or granulation tissue invades, trabeculae are destroyed, and healing takes place by fibrosis. This granulation tissue and infiltrating cells give a characteristic yellow color to the medulla. If the defenses are inadequate to cope with virulence of the organism, the tissues locally are destroyed, the necrosis producing a typical yellowish-gray gummatous detritus, the gumma. The central necrotic focus is surrounded by small round cells, outside of which granulation tissue and fibrous tissue are seen. Beyond the lesion, reactive new bone forms. The osteoblastic reaction is extensive. The medullary infective material extends through the haversian systems to the subperiosteal space where elevation of the periosteum results in successive deposits of laminated bone. If the infection infiltrates the periosteum without elevating it, the reactive bone assumes a lacelike appearance. Periosteal bone becomes incorporated within and thickens the cortical bone. Therefore, periosteal ossification is a natural consequence of (1) elevation of periosteum by exudate or granulation tissue, or (2) thickening of the periosteum by granulation tissue which organizes into fibrous tissue and subsequently undergoes osseous transformation. Granulation tissue, fibrosis and ossification imply that the infection is low-grade and defenses are adequate.

When the infection is virulent and defenses are inadequate, necrosis, destruction and gummatous formation of the periosteum and the subperiosteal space predominate, new bone formation being held to a minimum. The destruction erodes the cortex and may invade the medullary cavity In the skull, both cortices may be penetrated and the central nervous system infected. Or the infective material may burrow and erupt externally, forming a sinus discharging viscous, sticky material. Later, as a result of secondary infection, the discharge becomes purulent.

The joints are rarely primarily involved. The synovitis causing large painless effusions in late congenital syphilis or occasionally in acquired tertiary syphilis is due presumably to mild synovial infection, but this has never been proved. The synovitis is often bilateral, and the fluid is clear and sterile.

Gummatous osseous destruction of long bones is rare, but when it does occur it usually

[11] Campos, O P. Bone and joint tuberculosis; treatment, J. Bone & Joint Surg 37A:937, 1955.

prone to enlargement and deformity by osteoblastic changes, creating the markedly increased density seen in roentgenograms. The contour of the shaft is spindle-shaped, and the surface may display bone erosion. When multiple areas of gummatous destruction are present within the enlarged bulbous bone, the x-ray picture may be confused with that of tuberculous dactylitis. Clinically, the finger displays a large, spindle-shaped, boggy, painless swelling. Bilateral lesions are not uncommon.

Adult Syphilis

Bone and joint lesions occur in the tertiary stage years after the primary stage. At least 50 per cent of luetics have bone involvement, although a smaller number are manifest clinically and roentgenographically. The infection may be mild and resolve without apparent alteration of bony architecture. A slightly more virulent infection, assuming that the defenses are adequate, stimulates an *osteoblastic reaction,* the cortex becoming dense and thick and encroaching upon the medullary cavity. On the other hand, a more virulent infection, although continuing to stimulate surrounding osteoblastic reaction, is destructive. The resultant lesion is the *gumma.* The products of necrosis produces a yellowish gumlike material. The infective material may spread through the haversian canals to the subperiosteal space where further gummatous necrosis occurs. Or the medullary infection may progressively destroy the cortex, the periosteum and the soft tissues, finally erupting as a chronically draining sinus. Secondary infection is usual. *Sequestra are rare.* Involvement, is greatest at the ends of the long bones where the joint may be invaded, and a gummatous arthritis may result.

Periostitis is the most common lesion, occurring in both skull and long bones The periosteum becomes edematous and infiltrated with round cells and granulation tissue. The granulation tissue is converted to fibrous tissue which in turn is ossified by a lacelike deposition of new bone.

In the skull the outer table is chiefly involved, but gummatous destruction may erode through the inner table and produce a luetic meningitis.

Healing of gummata takes place by fibrous tissue replacement and conversion to bone. Marked thickening and sclerosis of the involved bone results.

Clinical Picture. Bone lesions become suspect because of a *painless* fixed, nontender swelling of a long bone or the skull; or a soft nontender, noninflammatory swelling may be revealed by roentgenograms as an extension from a gummatous osteomyelitis. *Luetic lesions are characterized by absence of pain and lack of acute inflammatory signs.* When the gummatous destruction invades a neighboring joint, considerable effusion, irregular lumpy swellings of the periarticular tissues, little restriction of joint motion, lack of muscle atrophy, and absence of pain are characteristic. If destruction is extensive, eruption externally and secondary infection produce a suppurative arthritis.

Roentgenographic Findings. The picture of early infection is negative. Prolonged infection is revealed by *extensive osteoblastic densities,* thickening of the cortex, and a lacelike or laminated subperiosteal new bone. *Gummatous destruction is manifest as a moth-eaten appearance* interspersed with areas of sclerosis. The entire shaft is thickened and may present an irregular or wavy surface. The x-ray appearance may be identical with chronic suppurative osteomyelitis.

Diagnosis

Syphilis has protean manifestations, affecting any organ of the body and simulating any disease. Similarly, it can resemble any bone and joint disease. The following characteristics of luetic infection of bone are important in establishing the diagnosis:

1. Lesions are predominantly osteoblastic.
2. When moth-eaten rarefactions of gummata are observed roentgenographically, they are almost invariably associated with extensive osteoblastic densities.
3. Tendency toward periosteal ossification (lacelike x-ray appearance) and thickening of the cortex.
4. Swellings on the external surface of the bone, whether hard (bone), indurated (periosteal proliferation), or soft (gumma), are painless, nontender, and lacking in inflammatory signs.

In the adjacent periosteum, periosteal involvement results in reactive new bone formation. The long bones, skull and nasal bones are favored sites of involvement.

If the infant survives the early days after birth, the condition shows a tendency toward healing within a few months. The bone infection clears up, and endochondral ossification is resumed. This suggests that the metaphyses are apparently resistant to infection.

Clinical Picture. Shortly after birth, the limb displays a large tender swelling about a joint. The child is irritable, restless and cries often. The limb is held immobile as though paralyzed. Other evidence of syphilis may be apparent, such as snuffles, keratitis, skin lesions, mucous patches, positive serology, etc.

Roentgenologic Findings. The metaphysis is widened, osteoporotic, or moth-eaten in appearance, and its epiphyseal border is dense, jagged and indented. The ossification center has not yet appeared, and it is difficult to ascertain the increased thickness or fracture displacements of the epiphysis Surrounding the metaphysis and extending toward the diaphysis, the laminations of periosteal ossification are seen.

Differential Diagnosis. Rickets and scurvy are the main conditions to be differentiated.

RICKETS. Syphilis occurs before 6 months of age, whereas rickets does not appear until after 6 months. In rickets, the provisory zone of calcification is demineralized and not apparent on roentgenographic examination. Instead, the epiphyseal plate appears to be widened. Periosteal ossification in rickets develops on the concave aspect of the long bone in response to static stresses, and deformity results from actual bowing of the bone. In lues, periosteal apposition of new bone has no relation to the concave aspect of the bone and is the actual cause of deformity, not bowing.

SCURVY. The epiphyseal plate is of normal width, but the bony structure of the metaphysis presents a ground-glass appearance The main changes occur about the circumference of the shaft. Vague densities represent subperiosteal hemorrhage which eventually becomes ossified.

Treatment. If syphilitic infection of the newborn is not extensive and the infant survives, the osseous involvement is self-limited and heals within several months. Nevertheless, antiluetic therapy brings about a rapid recovery, and interference with subsequent longitudinal growth is less likely.

LATE CONGENITAL SYPHILIS

This is usually regarded as the tertiary stage of congenital syphilis, occurring after the 2nd or the 3rd years. Osteoblastic changes characterize the lesions found mainly in the tibia, the femur and the skull. *Periosteal bone formation* occurs typically over the anterior aspect of the tibia, causing a bony prominence (saber shin). No actual bowing of the shaft takes place. In the skull nodular thickenings over the outer table produce deformity. Unless adequate antibiotic therapy is instituted, the periosteal bone is incorporated into the cortex as a permanent deformity. A more virulent subperiosteal infection may erode the cortex from without; or gummatous destruction of the cortex may arise from within the medulla. This destructive osteomyelitis is rare in congenital syphilis. The x-ray picture, whether destructive or not, displays the increased densities of osteoblastic changes.

Associated evidence of syphilis includes interstitial keratitis, 8th nerve deafness, and deformed incisor teeth (Hutchinson's triad). At least one fourth of syphilitic children have involvement of the central nervous system.

CLUTTON'S JOINTS

(Symmetrical Synovitis in Congenital Syphilis)

Large, bilateral, painless effusions of the knees, occurring in late congenital syphilis between the ages of 8 and 18, are designated as Clutton's joints. The condition arises spontaneously and intermittently without apparent cause and disappears as mysteriously. No local inflammatory or constitutional signs are present, and the many recurrences cause no damage to the joints. If the synovial fluid is aspirated, it rapidly reaccumulates Microscopic examination of the fluid reveals a high content of mononuclears. X-ray examination is negative.

SYPHILITIC DACTYLITIS

The phalanges and the metacarpals are

show small single buds being extruded externally. *Coccidioides* show central multiple spores resembling cocci. No new bone formation is seen.

Diagnosis

Material from an abscess is cultured in 10 to 40 days on blood agar or Sabouraud's medium. The culture colonies are placed in 10 per cent potassium hydroxide, which dissolves everything but the spores. Skin tests are performed with blastomycosis vaccine or coccidioidomycosis antigen. The complement-fixation test for *Blastomyces dermatitidis* is useful.

Treatment

The outlook is poor in the disseminated type. On the other hand, when a single focus exists, such as in a bone, cure is quite possible. The patient is desensitized with vaccine. At first, starting with a 1:100 dilution of vaccine, a subcutaneous injection is given 3 times weekly, the initial dose being 0.1 cc. and increasing the succeeding dose by 0.1 cc. until 1.0 cc. is reached. Then the same procedure is used with a 1:10 dilution. X-ray therapy is given on an empirical basis. Large doses of potassium iodide are administered. Healing is manifested by increase in density and regeneration of bone.

MADURAMYCOSIS

(Madurellamycosis; Madura Foot)

Maduramycosis is an uncommon mycotic infection involving bone and occurring in



Fig. 50. Madura foot.

5. The exudate from sinuses of gummata is thick and yellowish, but later, because of secondary infection, it becomes similar to pus from any osteomyelitic focus. However, sequestra are rare.

6. In congenital syphilis in infancy, infection is almost invariably confined to the metaphyses of long bones, and spontaneous healing before the 6th month usually occurs.

Serologic Tests. Various types of very sensitive serologic tests of the blood and the spinal fluid are now used to identify syphilis. It is important to note that an infant with *congenital syphilis cannot produce within the first* few months antibodies sufficient for serologic identification. Therefore, serologic reaction may remain negative until after the first 3 months. However, the diagnosis may be quickly ascertained after birth by quantitative serology. If antibody titer rises continually, the infant is regarded as a luetic. The question of diagnosing syphilis in a newborn makes it mandatory to examine both parents and children. The history of repeated spontaneous abortions in the mother, each one taking place in progressively later periods of gestation, is extremely suggestive of maternal syphilis.

In late syphilis, at the time when osseous involvement occurs, blood serology is not infrequently negative, and spinal fluid serology is positive. Therefore, examination of the spinal fluid should be done routinely and should include not only the serologic test but also a determination of protein content, cell type and count, and the colloidal gold curve.

MYCOTIC INFECTION OF BONE[12]

Mycotic infection of bone is rare. Blastomycosis and coccidioidomycosis are the usual fungous infections which destroy bone. The fungi reside in soil, and the disease is acquired by inhalation of spores. Reproduction occurs within the animal host: *Coccidioides* by endosporulation and rupture of its capsule; *Blastomyces* by budding externally.

The onset of the disease is insidious and causes a mild respiratory infection. Coccidioidomycosis, which is endemic in the Southwest, is the most prevalent of the two diseases. The coccidioidal respiratory infection is benign and subsides quickly. A few weeks later, an acute pneumonitis develops and heals with fibrosis, occasionally with cavitation. Most of these cases remain cured, but a very small percentage develop an "allergic reaction" within a few weeks characterized by fever, arthritis and muscle pains. The joints are tender, painful, and slightly swollen, but no effusion occurs, and symptoms subside completely. Nothing further develops as a rule, but a rare case develops systemic spread of the fungous infection. Granulomatous lesions can occur any*where. Bones, muscles, joints and skin are* often involved.

Blastomycosis is not endemic. After the original benign respiratory infection has subsided, within several weeks or months the patient begins to have a low-grade fever and loses weight and strength. A skin ulcer or a subcutaneous abscess is often the first symptom suggesting fungous infection. A subcutaneous nodule may break down, discharge a sanguinopurulent material and heal with a dense depressed scar, or it may persist as a chronic ulcer.

The bone lesions of blastomycosis and coccidioidomycosis are identical. The lesions arise in cancellous bone and are predominantly destructive with little periostitis, bone production, or marginal reaction. A well-defined area of osteolytic destruction is seen in roentgenograms.

PATHOLOGY[13]

The lesion is a granuloma. The tubercle may liquefy and form a chronic abscess and a sinus. Healing occurs by fibrosis and occasionally by bone reconstruction. Dissemination is by lymphatics and blood stream.

Microscopically, the appearance is granulomatous. The cells include histiocytes, epithelioid cells, polymorphonuclear leukocytes and lymphocytes. A fibroblastic reaction is present. Giant cells are present in moderate numbers. The small spherical bodies with double contoured capsules lie free in the tissues and within giant cells. They contain central clumps of basophilic material. Blastomyces

[12] Alfred, K. S., and Harbin, M.: Blastomycosis of bone, J. Bone & Joint Surg. 32A:887, 1950.

[13] Hall, R. H., and Mendelcff, J.: Blastomycotic osteomyelitis, J. Bone & Joint Surg. 34A:977, 1952.





Fig. 51. Sporotrichosis: Characteristics to be sought in identifying this cause of chronic granulomatous osteomyelitis, periostitis or synovitis. (*Top*) Typical primary lesion on dorsum of hand with multiple nodular and ulcerative secondary lesions arranged in linear fashion along a thickened lymphatic cord. (*Center*) *Sporotrichum schenckii* from culture showing tangled filamentous threads with short side branches upon which lie clusters of priform conidia. (*Bottom, left*) Tubercle formation. (*Bottom, right*) Infiltration of histiocytes and giant cells without necrosis. (Duran, R. J., Coventry, M. B., Weed, L. A., and Kierland, R. R.: Sporotrichosis, J. Bone & Joint Surg. 39A:1330)

tropical and subtropical zones and in the southernmost points of the United States, especially Texas. It is characterized by progressive local necrosis and abscess formation, intermittent discharge of a peculiar fluid through fistulous tracts, and replacement by granulation and fibrous tissue.[14-17]

Etiology

Individuals between the ages of 20 and 40 are affected. The mycelia are coarse and contain numerous round and oval bodies, the chlamydospores. They form grains or granules which vary in color but often are jet black. The mycelial threads are often arranged in clusters of radially arranged eosinophilic clubs The mode of transmission is unknown, but patients usually are agricultural workers giving a history of suffering from a thorn wound.

Clinical Picture

Maduramycosis predominantly affects the foot. The earliest stages of infection are rarely observed. The condition starts with multiple, hard, deep-seated and fixed papules or nodules which soften at their centers and form abscesses The abscesses rupture and produce persistently draining fistulae. The progress of extension is very slow, the infection spreading and deeply involving tendons, muscle and bone, and proximally toward the ankle and up the leg. Pain is minimal, and inflammatory signs are absent, so that the individual continues walking and furthering destruction. The big toe is often affected first. Involvement of bone invariably occurs with gross destruction and disorganization of all bones of the foot. The foot becomes swollen, indurated, misshapen and punctured with sinuses draining exudate containing varicolored granules but mainly black grains resembling caviar. Systemic reaction is absent, and the blood picture is normal. Infection and destruction is slowly progressive and persistent. The offending mycelia may be identified by culture and hanging-drop examination.

Roentgenographic Findings

These are extremely variable. The disease is most often observed at an advanced stage which exhibits extensive destruction of all bones of the foot. Rarely, a single lesion may be seen in the tibia where the picture is identical with chronic osteomyelitis.

Pathology

Microscopically, the dark grains are composed of coarse, dark mycelial elements, pigments and debris. A typical granulomatous reaction surrounds the abscess cavity, i.e., granulation tissue, round cells, giant cells and extensive scarring.

Differential Diagnosis

Leprosy, syphilis, tuberculosis, malignant neoplasm and other mycotic infection must be ruled out.

Treatment

Early bone lesions heal readily under the influence of sulfonamides and antibiotics Severe destructive lesions with draining sinuses require amputation.

SPOROTRICHOSIS OF BONE[18, 19]

Sporotrichosis is a fungus infection producing granulomatous lesions usually involving skin and subcutaneous tissue but uncommonly is disseminated systemically, involving various tissues and viscera. When bone is infected, the slow destructive and exudative process resembles chronic osteomyelitis or tuberculosis. The condition is being recognized with increasing frequency.

Etiology

The causative organism is *Sporotrichum schenckii*. It is almost never identified in secretions from the lesions or in biopsy material

[14] Gammel, J A.. Etiology of maduramycosis, Arch. Dermat & Syph 15 241, 1927 Correction, ibid. 15:477, 1927.

[15] Thompson, H L Present status of mycetoma, Arch Surg. 16 774, 1928

[16] Downing, J. G, and Conant, N F Medical progress: mycotic infections, New England J Med 233:153, 1945

[17] Kulowski, J, and Stovall, S Maduramycosis, J.A M.A 135:429, 1947.

[18] Conant, N. F, Smith, D T, Baker, R. D, Callaway, J. L, and Martin, D. S Manual of Clinical Mycology, ed 2, p 222, Philadelphia, Saunders, 1954

[19] Duran, R J, Coventry, M B, Weed, L A, and Kierland, R R.: Sporotrichosis, J Bone & Joint Surg. 39A 1330, 1957.

duce an acute and chronic osteomyelitis. The genus Salmonella, of which more than 150 types are known, are gram-negative bacilli possessing motility by means of flagellae. Three kinds of infection in man are identified: (1) gastroenteritis, occasionally severe enough to be fatal, (2) Salmonella fever, similar to typhoid fever, and (3) a septicemia characterized by multiple metastatic abscesses. The last-named type is often caused by *S. choleraesuis* which therefore is the most frequently isolated organism obtained from bone abscesses.[20, 21, 22]

Clinical Picture

This is similar to other types of acute osteomyelitis. Intestinal symptoms are usually absent. The infection shows a tendency to subside and persist as a chronic infection. The sites of predilection are at the ends of long bones and the lumbar spine. Multiple skeletal involvement may occur and end fatally.

Laboratory Findings

The causative organism may be cultured from the lesion, the blood stream and rarely from the stool. Agglutination tests are diagnostic. The organism may be traced to or isolated from certain foods.

Treatment

The organism is supposedly susceptible to chloramphenicol. The principles applied to other forms of osteomyelitis should be followed.

BRUCELLAR SPONDYLITIS[23, 24]

Brucella organisms occur in domestic animals and are transmitted to man by direct contact. *B. abortus*, found in cattle, is the most common organism infecting man. It tends to reside in the reticuloendothelial system for prolonged periods so that the course of the disease is protracted. A conglomeration of symptoms, often vague, include fever, sweats, weakness, generalized pains, headache, depression, etc. The examination may reveal nothing but fever, and half the cases have lymph node enlargement and a palpable spleen. *B. suis* (from swine) often causes suppurative osteomyelitis, usually in the vertebrae. *B. melitensis*



Fig. 52. Brucellosis of the spine. The 4th lumbar disk is destroyed, and the space is narrowed. The inferior cortex of the 4th lumbar vertebral body and the superior cortex of the 5th are destroyed and sclerotic. (Brucellosis, p. 22, Pfizer: Spectrum, Nov. 5, 1955)

[20] Guerra, R A, Pellufo, E, Laguarda, M, and Aleppo, P. L.: Septicemias and bacteremias in infants, Arch pediat Uruguay 10 669, 1939

[21] Ralston, E L: Osteomyelitis of the spine due to salmonella choleraesuis, J. Bone & Joint Surg 37A: 580, 1955.

[22] Weaver, J B, and Sherwood, L. Hematogenous osteomyelitis and pyarthroses due to salmonella suipestifer, J.A M A. 105 1188, 1935

[23] Feldman, W. H, and Olson, C Spondylitis of swine associated with bacteria of the brucella group, Arch Path. 16 195, 1933

[24] Mantle, J. A Brucellar spondylitis, J. Bone & Joint Surg 37B 456, 1955

but is seen best in a suspension of a culture. It appears as tangled, filamentous threads with short side branches upon which lie clusters of small oval bodies called conidia. The fungus exists in nature as a saprophyte of plants, flowers and trees, particularly in an environment of high humidity. Consequently, farmers, horticulturists, fruit-pickers and the like are favored victims of this occupational disease. The usual mode of infection is through a trivial wound in the skin, e.g., a thorn prick, but the disease can be transmitted by an animal bite or the handling of dressings of patients.

Pathology

The lesion is a granulomatous reaction with central area of necrosis surrounded by epithelioid cells, chronic inflammatory cells, and an occasional giant cell. Activity varies from destruction and exudation at one extreme to healing by resorption of exudate and fibrosis at the other. The granulomatous lesions in bones and joints are comparable with an extremely low-grade form of tuberculosis.

Clinical Picture

The initial lesion most often involves the upper extremity, usually the hand A history is often elicited of a specific trauma, e.g., pricking the finger on a rose thorn a number of days or weeks previously At this site an indurated, nontender, freely movable nodule develops and is covered with reddened skin Later, the color becomes violaceous, the nodule becomes adherent to the deep tissues and the skin, then softens and becomes fluctuant, and the overlying skin breaks down to form an indolent, granulomatous ulcer which repeatedly bleeds and crusts and heals very slowly over weeks to months Within a few days to a week after appearance of the initial lesion, infection spreads through and thickens lymphatic channels in the forearm, and these are palpable as thick cords. Along a prominent cord, a chain of nodules characteristically form, and these too break down, forming ulcers which discharge a watery exudate and persist for months or years. The general health is usually unaffected in this lymphatic form.

Uncommonly, systemic involvement occurs, with or without a recognizable antecedent lesion on the extremity. Granulomatous lesions can involve any tissue, supposedly by hematogenous spread, but pulmonary and central nervous system involvement is rare. Bone infection may appear as an osteomyelitis or periostitis, and synovial infection is not unusual. In addition to widely spread subcutaneous nodules, a generalized lymph gland enlargement is often apparent. A low-grade fever and slight leukocytosis with eosinophilia are often associated. Systemic sporotrichosis is a chronic, progressive illness with a grave outlook.

Diagnosis

The fungus cannot be identified in secretions or biopsy material. Instead, the biopsy tissue is ground up and cultured, and typical colonies are produced. A suspension of the growth reveals the fungus. Complement-fixation tests, agglutination tests and skin tests are not specific.

If an externally draining ulceration is found to communicate with subjacent infected and destroyed bone, secondary infection by bacterial contaminants makes this unsuitable for culture. Instead, a nodule or an enlarged lymph gland should be used

Sporotrichosis must be differentiated from tuberculosis, syphilis, pyogenic infection, tularemia, coccidioidomycosis, blastomycosis, histoplasmosis, squamous cell epithelioma, and granulomata caused by drugs.

Treatment

Potassium iodide is the drug of choice, especially for the localized lymphatic form. The dosage is gradually increased up to 50 mg. per day. The skin lesions readily heal, but treatment must be continued another 6 weeks to preclude recurrence.

For systemic sporotrichosis, 2-hydroxystilbamidine is given slowly intravenously in daily 225 mg. doses Dangerous toxic drug reactions are frequent.

The problems of handling bone and joint involvement are similar to those of tuberculosis.

SALMONELLA OSTEOMYELITIS

Salmonellae organisms which most commonly cause food-poisoning will rarely pro-

9

Congenital Deformities

A. Etiology

The actual cause of a deformity found at birth is theoretical. However, the two known factors are (1) *genetic* and (2) *embryonic trauma*.

GENETIC FACTOR

The genes in the chromosomes of the ovum and the sperm transmit the specific anomalous characteristic. Transmission follows Mendel's Law. When the genetic factors are dominant, the anomaly will develop in a large number of offspring. When the factors are recessive, the anomaly occurs infrequently. For example, osteogenesis imperfecta follows a pattern similar to hemophilia in that the disease occurs almost exclusively in the male but is transmitted by the female, as a mendelian recessive.

FACTOR OF EMBRYONIC TRAUMA

Experimental and clinical evidence has shown that many things can injure the developing embryo. In the early weeks of differentiation of the embryonic tissues into specific tissues and component parts of the fetus, the embryo is most susceptible to extraneous factors. Each component part rapidly differentiates at a specific time when it is most sensitive to trauma. Also, each injurious factor seems to have an affinity for a particular area or tissue. Therefore, the type of deformity might be determined theoretically by the traumatic agent and the time at which it exerts its deleterious influence.

TERATOGENESIS[1]

This is the experimental production of congenital anomalies. The following are the known teratogenic factors:

1. *Metabolic:* Hypoglycemia due to various disorders, including hyperinsulinism.
2. *Endocrine:* Adrenocoticotropic hormone (ACTH) or cortisone injected experimentally in animals or therapeutically in humans. Clubfoot is a common deformity resulting when such treatment continues during pregnancy.
3. *Nutritional Deficiency:* Particularly, lack of riboflavin. It is interesting to note that experimental production of deformities by insulin can be largely prevented by injecting nicotinamide and riboflavin at the same time as the insulin.
4. *Chemical:* For example, lead nitrate can induce hydrocephalus, meningo-encephaloceles and meningomyeloceles in chick embryos.
5. *X-radiation:* A cleft palate can be produced experimentally.
6. *Infection:* The frequent association of cataracts and cardiac septal defects with maternal intercurrent infection of rubella during the early months of pregnancy strongly suggests the cause and the effect.
7. *Mechanical:* Injury by direct mechanical trauma to the embryo in the early weeks of pregnancy must be considered in the light of experimental evidence. For example, "rumplessness" can be produced in chickens by shaking of the eggs.
8. *Thermal:* The earliest postconceptional phase is the most susceptible time in experimental animals.
9. *Anoxia:* Anencephaly, spina bifida and other defects produced in laboratory animals suggests an important clinical application in pregnancy maternal anoxia of anemia.

[1] Duraiswami, P. K.: Experimental causation of congenital skeletal defects in orthopaedic surgery, J. Bone & Joint Surg. 34B.646, 1952.

causes severe general symptoms and neurologic disturbances such as peripheral neuritis, cranial nerve palsies and meningo-encephalitis.

The spine is frequently infected by brucellar organisms. The pathology consists of mononuclear infiltration, bone destruction and abscess formation. The abscesses have tough fibrous capsules. Occasionally, sequestra are formed, but early and extensive new bone formation is invariable. The abscesses extend to the dura but do not penetrate it. The intervertebral disk is often destroyed, and bony fusion between adjacent vertebrae may follow.

Symptoms in man include fever, pain which is often severe, and rigidity of the spine. Deformity is rare. The course is more rapid than tuberculosis. The temperature is intermittently elevated, and the sedimentation rate is high. Paraplegia is rare.

X-ray studies show early and extensive bone proliferation, absence of rarefaction, early sclerosis of the bodies, and narrowing of the disks. Often bony fusion of the vertebrae is observed. The picture may be confused with tuberculosis of the spine.

Diagnosis is mainly by agglutination tests and blood culture. Sternal marrow biopsy may reveal the typical lesions and organisms.

Treatment

Chlortetracycline is very effective. Oxytetracycline, streptomycin and sulfadiazine are also beneficial. Spine fusion may be necessary to eliminate pain.

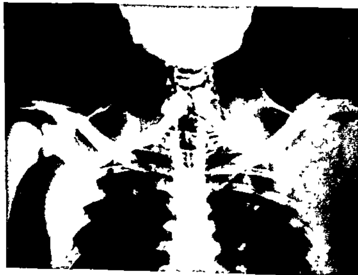
veloping muscles (supraspinatus, infraspinatus, subscapularis) and those which attach to the scapular spine and the vertebral border (trapezius, levator scapulae, rhomboids, serratus anterior) are composed of normal muscle tissue, or imperfectly developed muscle fibers having the appearance of myoblasts, or wholly of fibrous tissue. From the superior angle, a sheet or bandlike structure composed of fibrous tissue, cartilage, or bone extends upward to attach to the transverse processes of several cervical vertebrae. Occasionally, it reaches the base of the occiput of the skull. This structure probably represents the levator scapulae. Associated errors in segmentation are represented by wedged and fused cervical and thoracic vertebrae, hemivertebrae and fused ribs, all of which are responsible for the frequently associated scoliosis. Congenital failure of fusion is displayed by the spina bifida in the cervical spine and the separation of the occipital bones extending upward from the foramen magnum.

Clinical Appearance

As viewed from the rear, the shoulder and the scapula are higher than on the opposite side. When the condition is bilateral, the upward displacement of both shoulders gives the neck a shortened appearance. The scapula is rotated counterclockwise, the inferior angle displacing toward the spine. At the upper angle of the scapula, the band which extends up toward the cervical spine or even the base of the skull may be apparent as an elevated ridge. Upon attempted abduction of the arm, the lower angle of the scapula does not rotate outward as it should in normal scapulohu-



Fig. 53. (*Right*) Sprengel's deformity, or congenital elevation of the scapula. (*Bottom*) Bilateral undescended scapulae. The roentgenogram reveals persistence of the embryonic connection with the cervical spine, the omovertebral bone, on each side. Clinically, the omovertebral bones form prominences extending from the cervical spine to the superior angle of the scapulae. (U. S. Army photograph. Col. Blair, J. D., and Wells, Col. P. O.: Bilateral undescended scapula associated with omovertebral bone, J. Bone & Joint Surg. 39A:201)





10. *Maternal Iso-immunization:* Antigens present in the blood of the fetus (inherited from the father), though lacking in the mother's blood, produce maternal Rh iso-immunization which may interfere with normal fetal development and result in extensive abnormalities of the central nervous system. The following are the features: severe mental deficiency, asymmetric hypertonicity and weakness of the extremities, the neck and the back, and choreo-athetosis. In almost all cases it is possible to obtain a neonatal history compatible with *erythroblastosis fetalis* and an Rh setup in the mother and the child compatible with the diagnosis.[2]

Duraiswami, after his classic experiments in experimental production of skeletal defects, made the following summarizing hypothesis:

> The development of an embryo, which is presumably guided by a series of organizing processes, may be interfered with during critical periods by genetic or environmental teratogenic factors Such disturbances caused to the organizer system may, in their turn, produce metabolic, biochemical, and other changes through the intervention of hormones and enzyme systems and thus interfere with the normal development of the embryo. The resulting abnormalities in development would not occur in a random assortment, but tend to fall into certain categories corresponding to the critical stages of development of susceptible tissues and the quality and intensity of the noxious agent. Developmental malformations may result not only from arrest of growth and differentiation of the embryo as a whole, or some of its parts, but also from degeneration in tissues which had developed normally up to a certain stage, as has clearly been demonstrated in the case of insulin induced deformities and rubella induced lenticular lesions. It is possible to demarcate in the life of the human embryo certain critical periods which are peculiarly associated with catastrophic changes in the development of the skeleton. The fourth and fifth weeks of intra-uterine life are definitely associated with the development of the cartilage skeleton. The seventh and ninth weeks present widespread calcification of the cartilage of the long bones as the main feature. The last two weeks of fetal life are marked by the rapid growth in length of the long bones, as distinct from the increase in size of the epiphyses. The orderly progression from the mesenchymatous condensation to cartilage, and then through calcified cartilage to bone may be disturbed by genetic or environmental teratogenic factors and a variety of skeletal deformities result.

The following sections describe conditions more commonly seen and treated in orthopaedic practice.

[2] Yannet, H: Mental deficiency due to prenatally determined factors, Pediatrics 5 328, 1950

B. Upper Extremity

CONGENITAL HIGH SCAPULA

(Sprengel's Deformity, Undescended Scapula; Elevated Scapula)

This deformity consists of a permanent elevation of the shoulder girdle

Etiology

Like other congenital deformities, the unknown causative factor is operative during early embryonic life, but most particularly at the time of development of the cervical spine and the upper limb buds and subsequent descent of the latter. This explains the frequent association of deformities of the occiput and the base of the skull, the cervical and the upper thoracic spine, the ribs and the surrounding muscle tissues. Before the third month, the mesenchymal tissues take form as the cervical spine and, at the same level, the upper limb buds. The limb buds then descend to the level of the thorax. The failure of descent results in a permanently high shoulder girdle and imperfect development of the surrounding tissues, described as incomplete segmentation and the failure of fusion of bony elements.

Pathology

The scapula is smaller in its vertical diameter so as to appear broad. The suprascapular portion arches forward where it fits over the superior thoracic cage in its elevated position. Looking at it from the posterior aspect, it is rotated counterclockwise so that its inferior angle is approximated to the spine. The en-

going the extensive surgery necessary for correction. When function at the shoulder is restricted, restoration of position to the scapula does not improve function. Correction should be undertaken in childhood. The downward displacement may cause traction on the brachial plexus, and the nerve trunks may be compressed by lowering the clavicle which thereby narrows the costoclavicular space.

Surgical Procedure. A longitudinal incision is made along the vertebral border of the scapula. The attachments of the muscles to the vertebral edge are incised. The rhomboids are reflected toward the spine; the supraspinatus and the infraspinatus are elevated laterally; the trapezius is cut from its attachment to the spine of the scapula; the subscapularis is freed from the undersurface; the serratus magnus is removed from the medial border; and the fibrous or chondrous or osseous band is cut from the upper angle and removed. In exposing the band, the posterior scapular artery should be sought and ligated. The suprascapular nerve and artery are identified and protected at the suprascapular notch. The completely denuded scapula is now freed for displacement except for the clavicle, which prevents descent. A Z-plastic osteotomy of the clavicle permits lengthening of the bone and downward movement.

An alternative is removal of the outer end of the clavicle, including the conoid and the trapezoid ligaments which attach to the coracoid process of the scapula. A wire suture is inserted through 2 drill holes in the scapula and directed downward to emerge through the skin distally. This wire will be fixed to a body spica cast under tension to maintain the lower position of the scapula. A number of muscle fibers may be freed from the erector spinae from below and attached to the inferior angle; supposedly, this aids in maintaining the new position. Fixation of the lower angle to a rib is not recommended, as this may restrict scapular motion. The wire is removed when the clavicle has united.

CLEIDOCRANIAL DYSOSTOSIS[3,4]

Cleidocranial dysostosis is a congenital developmental condition in which membranous bones fail to ossify sufficiently, particularly in the calvarium and the clavicles, where fibrous tissue replaces the bone. Characteristically, the pubic bone also participates in the pathologic process, but the replacing tissue gives rigidity and strength to the pelvis which therefore does not feel deficient to the examining finger. Muscle deficiencies occur at the shoulder.

Clinical Picture

The following are the characteristics:

1. *Slender build, large head* with small shrunken face, long neck, drooping shoulders and narrow chest.

2. *Skull* shows well-marked bosses over the frontal, the parietal and the occipital areas. A median groove separates the frontal prominences. The anterior fontanelle is large and may never close completely. A mild degree of hydrocephalus may be present. The maxilla is small, and the relatively large mandible may appear prognathous. Delayed or deficient dentition is common.

3. *Clavicles* may have a defect at the inner, the middle or the outer third, or may be altogether devoid of bone. The remaining bone segment, particularly at the outer third, may compress the underlying nerves of the brachial plexus. The shoulders droop and can be approximated voluntarily. The bone defect is usually bilateral. Absence of the clavicular portion of the trapezius and the anterior fibers of the deltoid may occur occasionally. However, impairment of shoulder function is unusual. The scapulae may be small, deformed and winged.

4. *Widespread spina bifida occulta* is common.

5. *Deficient ossification of the pubic bones.* This causes no clinical impairment.

Roentgenographic Characteristics

1. Membranous calvarium imperfectly ossified.
2. Base of skull normally ossified.
3. Sutures often fail to close normally.
4. Anterior fontanelle large, may never close.

[3] Fairbank, H. A. Atlas of General Affections of the Skeleton, Baltimore, Williams & Wilkins, 1951.

[4] Marie, P. S, and Sainton, P.: Observation d'Hydrocephalie Hereditaire (Pere et Fils) Pare Vice de Developpement du Crane et du Cerveau, Bull et mém. Soc méd. hôp. Paris 14:706, 1897.



FIG. 54. Cleidocranial dysostosis.

meral rhythm. The amount of restriction of shoulder movement is variable and generally proportionate to the pathologic replacement of periscapular muscle tissue, and the presence of the superior restricting band. Very often the range of motion and strength is surprisingly excellent. Cervicothoracic scoliosis and torticollis are frequently associated features. Pain is absent.

TREATMENT

This is undertaken for cosmetic reasons only. There is no other justification for under-

FIG. 55. Cleidocranial dysostosis (*Top*) Typical pear-shaped brachycephalic skull. Note enlarged anterior fontanel, through which can be seen persistent posterior fontanel; also wormian bones about lambdoidal suture. (*Center*) Absence of outer portions of both clavicles, coracoid processes and supraspinous fossae of both scapulae. (*Bottom*) Wide separation of symphysis and acetabulum and irregular ossification of capital femoral epiphyses and acetabulae (Eisen, D.: Cleidocranial dysostosis, Radiology 61:21)

finger and the volar aspect of the other, and the flaps are brought over to cover the defects. Rarely are the flaps of sufficient length, and the resulting scar tissue formation jeopardizes any future surgery.

2. **Free Full-Thickness Graft.** This is the most successful procedure. The web is split, and a free graft, including skin to cover the commissure, is placed. A pressure bandage and splint are applied. If more than 2 fingers are involved, only one side of a finger at a time is operated to avoid bilateral compression of the finger which might compromise the circulation. If only one common tendon is found, it is detached from the poorer finger and left attached to the better developed one. Later a tendon may be grafted.

Symphalangism

This consists of fusion of interphalangeal joints, usually the middle with the proximal or distal phalanx. It occasionally accompanies syndactylism. Treatment is by arthrodesing in the functional position of partial flexion. However, if the symphalangism involves only the proximal joint and the distal one is moved by an adequately functioning profundus tendon, arthroplasty may be attempted.

Polydactylism

Reduplication of fingers varies from a completely developed finger to the doubling of a single phalanx. There is a tendency for the accessory digit to be located on the radial or ulnar margin of the hand. The types include (1) *an extra fleshy mass* which is not adherent to the skeleton and is devoid of bones, cartilage or tendons; (2) *duplication of a digit* which is composed of normal components and articulates with an enlarged or bifid metacarpal; and (3) *a digit with its own metacarpal*, which is rare.

Treatment consists of removal. One must ascertain whether the remaining digits have functioning tendons, because it may be necessary to transfer the tendon of the accessory digit, for example, to the thumb Reduplication of the distal phalanx is most common in the thumb (bifid thumb). If one segment is well developed, the other may be removed. Usually, it is better to excise a central wedge from each tip and join the remaining segments (Bilhaut-Cloquet Operation).

Brachydactylism

Shortening of the fingers due to decreased length of phalanges, decreased number of phalanges, or shortened metacarpals is a hereditary condition transmitted as a simple mendelian dominant. Frequently, it is associated with webbed fingers or polydactylism. The fingers may have good profundus tendons and function well. On the other hand, the tendon is absent, and the distal joint is flail or is a fused nonfunctioning joint. A flail distal joint should be fused to improve the function of pinch and grasp.

Annular Grooves and Congenital Amputations

Congenital constricting bands vary from a shallow groove in the skin or the subcutaneous tissue in one finger to a deep groove almost to the bone with a small distal segment of finger attached by a pedicle. Usually, it is associated with syndactylism and brachydactylism. The shallow ring does not interfere with function, but deeper rings cause distal edema.

Treatment consists of excising the grooves down to normal structures, approximating the subcutaneous tissue, and closing the skin by a zigzag plasty to avoid a constricting annular scar. Only one half the circumference of the finger should be operated at a time. Complete congenital amputation of fingers or hand is regarded as the extreme degree of congenital constricting ring.

Cleft Hand or Lobster-Claw Hand

This anomaly is extremely rare. A defect exists in the central portion of the hand. A V-shaped cleft tapering proximally may divide the hand into two parts, each part consisting of webbed fingers; the middle finger and the metacarpal are missing; no true thumb is present; grasping is accomplished by approximating the claws. In another type the middle finger is absent, but the metacarpal is present, and the cleft is shallow. The third type consists of a radial digit or thumb and an ulnar digit, the other fingers and the metacarpals being absent.



FIG. 56. Syndactylism. The skin bridge between the index and the middle fingers is complete, and that between the other fingers is incomplete.

5. Wormian bones in occipital and posterior parietal regions.

6. Maxillae are hypoplastic; mandible of normal size.

7. Partial or complete defect of clavicles.

8. Bilateral deficient ossification of pubic bones.

9. Congenital coxa vara is a frequently associated anomaly.

10. Failure of fusion of neural arches is common.

11. Epiphyses found at both ends of metacarpals and metatarsals.

TREATMENT

No treatment is necessary, since no disability is present. These patients live normal useful lives and have a normal age expectancy.

CONGENITAL ANOMALIES OF THE HAND[5]

ETIOLOGY

The accepted theories are those of maldevelopment or mutations which are subsequently inherited A mutation is defined as a permanent transmissible change in the character of an offspring from that of his parents.

SYNDACTYLISM

This anomaly is defined as joined fingers It is the most common deformity of the hand and is most frequent between the middle and the ring fingers and between the second and the third toes. It is often associated with polydactylism. Occasionally, hypodactylism and deficiencies of the long bones are accompanying anomalies. Experimentally, a diet deficient in riboflavin produces shortened or absent bones and syndactylism. The degree of syndactylism varies from 2 otherwise normal fingers being joined by skin (webbing) to the similar webbing between all the fingers (mitten hand). The webbing may be shallow or involve the entire length of the digits. The extreme degree involves fusing between the bones and joining of tendons and nerves. Therefore, it is essential that adequate preoperative x-ray study be made and careful deliberate dissection at surgery be enforced.

Treatment

Treatment is surgical. The skin of the web is rarely sufficient for covering defects when the fingers are separated. A free full-thickness graft is usually necessary. The following are the generally accepted procedures:

1. **Flap Operation (Didot Procedure).** At the area corresponding to the web, a triangular flap of skin with the base proximal is cut. It is routed through an opening cut on the volar aspect and is sutured to the palmar skin. The tunnel is kept open during healing by a wax stent or a glass rod. Later, an incision is made longitudinally over the dorsal aspect of one

[5] Barsky, A. J: Congenital anomalies of the hand, J. Bone & Joint Surg. 33A 35, 1951

finger and the volar aspect of the other, and the flaps are brought over to cover the defects. Rarely are the flaps of sufficient length, and the resulting scar tissue formation jeopardizes any future surgery.

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Treatment. In attempting to treat this condition, one should remember that any procedure which eliminates the cleft probably will reduce the function. The ability to grasp is accomplished by approximating the claws over an object held in the cleft. On the other hand, deepening a shallow cleft will improve function. When one considers amputation for unsightliness, the factor of excellent function of the lobster-claw hand as compared with the artificial hand should countermand the procedure.

The Thumb-Clutched Hand[6]

When one or both of the extensors, especially the long extensor, of the thumb are absent, the thumb is bent into the palm by the overacting flexors. A tendon transfer such as one of the extensors of the index finger will restore normal appearance and function.

[6] White, J. W., and Jensen, W. E.: The thumb-clutched hand, J. Bone & Joint Surg. 34A:680, 1952.

Megalodactylism or Macrodactylism

True congenital hypertrophy of the digits or hand is rare. The second or the third finger is usually involved. If unsightly, and particularly if unnecessary to hand function, it may be amputated. More frequently an enlarged finger is due to a local process such as a neurofibroma, an angioma, and another type of tumor which when removed improves appearance.

CONGENITAL ABSENCE OF THE RADIUS

This is a common congenital anomaly. It is usually associated with a clubhand which is

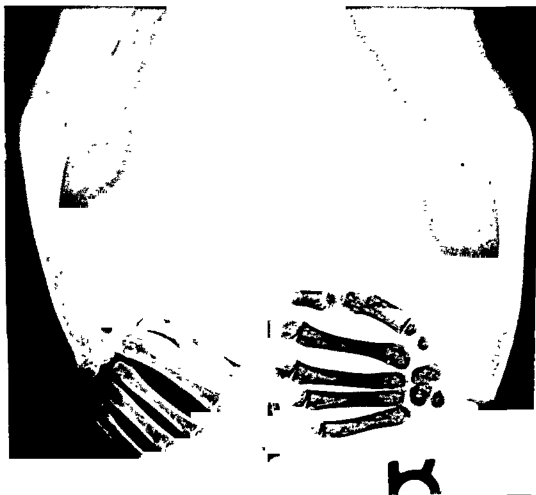


Fig. 57. Congenital absence of radius Deformity corrected by ulnarmetacarpal arthrodesis. (Case of Dr. D. Miller)

deviated radially. The bone may be completely absent, or a portion, usually at its upper end, may remain. The hand lacking the support of the radius at the wrist deviates radially until it lies at right angles to the long axis of the forearm. The ulna gradually bows in following the hand with the concavity on the radial side. It is usually short and thickened. The radial carpal bones are absent or fused. A contracture exists on the radial side of the forearm in the form of muscles which are shortened, fibrotic, fused to each other, or absent. These are the brachioradialis and the muscles to the radial side of the wrist, the thumb and the index finger.

Treatment

First, the contracture must be released. In mild deformity, this can be accomplished by stretching and splinting. In the severe type, surgical release is necessary. The tendons are lengthened or, where their absolute lack of function can be determined, they are severed; the fascia is cut transversely; the deformity is not straightened immediately for fear of cutting off the circulation through the radial artery. Gradual stretching and splinting secure the correction, if necessary by addition of an osteotomy of the ulna. Finally, the wrist is arthrodesed in the position of function.

CONGENITAL RADIO-ULNAR SYNOSTOSIS

The term "primary" radio-ulnar synostosis is applied to the congenital condition in contrast with "secondary" or traumatic type. Primary synostosis is usually bilateral and at the upper third of the forearm, although the junction can occur anywhere. The marrow cavity of the synostosis may be continuous with that of both bones. The upper end of the radius may or may not be perfectly formed, and anterior or posterior dislocation at the elbow is not unusual. The supinator brevis is absent and may be replaced by bone. Other muscles concerned with rotation of the forearm, such as the pronator teres and the quadratus, are imperfectly formed or absent. The shaft of the radius crosses over the ulna in close relationship to the latter so that a fixed pronation of the forearm and a tightened, narrow interosseous membrane result.

Treatment

Treatment is limited to osteotomy to place the forearm in the mid-position for better function. Attempts to overcome the synostosis and give rotatory function to the forearm are doomed to failure because of lack of properly functioning muscles. Freeing of the radius requires extensive surgery, which is not justified by results. Briefly, the procedures which have been attempted consist of:

1. Resection of the synostosis.
2. Resection of the head of the radius; cover stump with fascia lata.
3. Loop of fascia lata about the shaft of the radius and attached to a drill hole in the ulna to keep the radius from displacing.
4. Sever the interosseous membrane along much of its length.
5. Osteotomize radius and rotate to mid-supination or full supination.
6. Tendon transfer to radius to effect active supination.

CONGENITAL HUMERORADIAL SYNOSTOSIS

Like radio-ulnar synostosis, this is an error in segmentation in embryonic life. Therefore, very commonly it is associated with other synostoses throughout the forearm and the hand. When a joint is nonexistent, the muscles which would ordinarily move that joint are absent or imperfect. Therefore, it is unwise to attempt an arthroplasty unless adequate muscle for transfer is available. In this condition, if the arm-forearm angle is nonfunctional, the position should be corrected by an osteotomy. The lack of epiphyses at the lower humerus and the upper radius may cause some reduction in longitudinal growth, whereas the ulna would continue at its normal rate, resulting in a bowing deformity. This can be corrected by osteotomy. Fortunately, most of the longitudinal growth in the upper extremity takes place at the upper humerus and the lower radius and ulna so that eventual length is not reduced seriously.

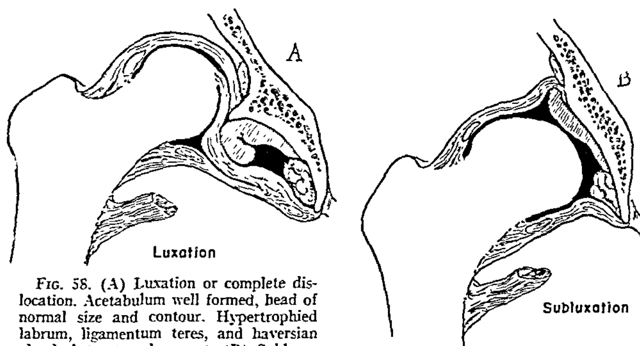


FIG. 58. (A) Luxation or complete dislocation. Acetabulum well formed, head of normal size and contour. Hypertrophied labrum, ligamentum teres, and haversian gland obstruct replacement. (B) Subluxation or incomplete dislocation Head enlarged and oblique. Acetabulum shallow. Roof inadequate. Labrum and haversian gland thin.

C. Lower Extremity

CONGENITAL DISLOCATION OF THE HIP

Displacement of the femoral head from its normal position within the acetabulum found at birth is regarded as a congenital dislocation of the hip. This must be distinguished from dislocations due to trauma, paralysis or infection occurring after birth.

ETIOLOGY

The actual cause is unknown. The hereditary factor is pronounced, and involvement of several members of one family is not unusual. Females are most commonly affected in the ratio of 9:1. The condition is prevalent in Mediterranean countries, notably Italy, where whole clinics may be devoted to the care of such cases.

PATHOLOGIC ANATOMY

Anatomic studies of the human fetus have demonstrated that at 10 weeks the femoral head, the acetabulum, and the capsule are well developed and proportionate as in the adult.[7] Therefore, the cause is operative, and the pathology develops before the 10th week in the anlage of the hip. At birth, the acetabulum and the femoral head are entirely cartilaginous; the antero-inferior acetabulum is shallow, and the cartilage is thin; the labrum glenoidale and the acetabular rim are well developed; anteversion of the femoral neck is 20° to 40°; inclination of the femoral neck is 110° to 120°; and extension at the hip is limited to 160°, the range of motion improving when the child starts walking. Deviations from this basic picture are associated with displacement of the femoral head.

The femoral head may be partially displaced upward (subluxated) or completely dislocated out of the acetabulum. The clinical picture and the treatment as well as the pathology varies with the two situations.

SUBLUXATION

The acetabular fossa is shallow and small, and the roof or superior portion is oblique, even vertical, offering no resistance to the upward glide of the head by muscle pull or weight-bearing. The superior acetabular pole is grooved and irregular due to constant friction, and at this point the labrum and the

[7] Howorth, M. B · Congenital dislocation of the hip, Ann. Surg. 125 216, 1947.

reflected tendon of the rectus femoris are pushed against the ilium and attenuated. The acetabular fossa assumes an oval or triangular shape. In comparison with the small size of the cavity, the femoral head is enlarged and cannot be adapted to the inadequate socket. The capsule is thickened, and its cavity is enlarged to accommodate the movement of its contents. The ligamentum teres may be elongated, hypertrophied, degenerate, attenuated, or absent. The anteversion of the femoral neck may be increased. The inferior and central portion of the acetabular fossa may be filled with fibrofatty tissue. The picture suggests atrophy of the socket due to lack of normal pressure by the head of the femur. When the head is replaced, especially early while the structures are mainly cartilaginous, the cavity will reform.



FIG. 59. Complete dislocation of the hip. (*Top*) Anteroposterior view shows an apparently foreshortened neck which is in reality a forward pointing bone. The acetabulum is somewhat inadequate. (*Bottom*) The anteversion is readily demonstrable in the lateral view.

COMPLETE DISLOCATION

Here the femoral head is completely displaced out of the acetabulum and comes to rest against the lateral wall of the ilium. Howorth states that most commonly it lies anteriorly adjacent to the antero-inferior iliac spine and secondarily comes to lie in the posterior area near the sciatic foramen. The pressure of the head against the ilium causes the former to be somewhat flattened posteriorly and the femoral neck to increase its anteversion. The capsule is greatly hypertrophied and increased in extent, and frequently this laxity of the capsule has been blamed for the dislocation. However, it is well known that a joint capsule will stretch from abnormal pressures within it and conversely will contract adaptively when intracapsular pressures are absent. As the capsule is pulled upward, it drags up the transverse ligament, and both capsule and ligament become adherent to the floor of the fossa and obstruct replacement. The femoral head pushes the capsule above the superior acetabular lip, and the capsule becomes adherent to the ilium. Pressure of the head against the ilium at the supra-acetabular area causes the capsule and the periosteum to differentiate into a fibrocartilaginous tissue lining a depression in the bone, thus forming a secondary socket or false acetabulum. Below this, the reflected rectus tendon and the glenoidal labrum obstruct replacement. The ligamentum teres may be elongated and markedly hypertrophied and in itself may interfere with reduction. However, it may also be attenuated or absent. The haversian gland and a thick labrum glenoidale fill the socket which, however, is deep and well developed. The iliopsoas tendon crosses the capsule and gives the latter an hour-glass appearance. The muscles which originate on the pelvis and insert distally as well as the fascia lata are adaptively shortened and prevent distal replacement of the femur.

CLINICAL PICTURE

The normal infant displays folds in the groins, below the buttocks, and several along the thighs. These folds are symmetrical. In hip subluxation and dislocation these folds are asymmetrical. As the infant lies on the examining table, the pelvis and the limb on the affected side are pulled proximally by muscle action as though the ipsolateral abdominal and



FIG. 60 The Allis sign. The knee is lower on the dislocated side.

spinal muscles were overactive as well as the hip adductors. Indeed, this factor should be considered in the etiology. The proximal displacement of the limb causes an apparent shortening which is never as pronounced by actual measurement, except in actual dislocation.

In the subluxated hip, when applying traction or testing abduction, a palpable or audible click is discernible as the head slips back and forth over the acetabular rim, and abduction is limited. When the hips of the normal infant are flexed to 90° and the thighs are abducted, the latter will nearly reach the table. In subluxation or dislocation, the abduction is restricted.

In a complete dislocation, the findings are more prominent. The femoral head may be palpable below the anterior superior iliac spine or posteriorly at the sciatic notch The shortening of the extremity is definite by actual measurement. The limb can be displaced by pushing and pulling, the femoral head being palpated as it moves to and fro. This is known as *telescoping*. The trochanter is prominent on the affected side.

When both hips are involved, the prominent trochanters give a widened appearance to the pelvis, and the lateral displacement of the thighs from each other causes a widened perineum. The child is late in walking

In unilateral dislocation, the typical gluteus medius gait is apparent. Because the femoral head floats freely, the gluteus medius has lost its fulcrum and cannot perform its function of elevating the opposite side of the pelvis as the opposite extremity is lifted off the ground. Compensation is obtained by swaying the body toward the affected side so that the center of gravity is thrust over the femur. This gait is found also in other conditions with gluteus medius deficiency, e g., poliomyelitis. When both hips are dislocated, the swaying from side to side produces the characteristic "duck-waddle" gait. The *Trendelenburg sign* tests the efficiency of the gluteus medius. Normally, when the lower extremity is lifted from the ground, the pelvis on that side rises due to contraction of the contralateral gluteus medius. If the pelvis sags downward, the sign is positive, and the muscle is regarded as inadequate. Looking at the patient from the side, the lumbar spine is extremely lordotic, and the abdomen is protuberant as the pelvis tilts forward when a bilateral dislocation exists and the femoral heads lie posteriorly. When the dislocations are anterior, the converse is true, i.e., the pelvis is horizontal, and the lumbar spine is flattened. A unilateral dislocation can be demonstrated by the *Allis sign*. With the infant lying on his back, the knees are flexed, and the feet are resting on the table. The knee of the affected limb will lie at a lower level.

Normally, walking starts at 11 or 12 months In this condition, however, walking is late. Older children will complain of pain, weakness and fatigability. Occasionally, the condition surprisingly occasions little complaint. In cases of subluxation, osteoarthritic symptoms eventually appear. Therefore, it is extremely important to recognize the earliest signs, particularly limitation of hip joint abduction (due particularly to contracture of adductor muscles), asymmetry of thigh folds, and slight or apparent shortening. X-ray studies are invaluable in concluding the diagnosis.[8]

ROENTGENOGRAPHIC STUDIES

At birth the acetabulum and the upper femoral epiphysis are cartilaginous and therefore not visualized in roentgenograms. Complete dislocations are easily detected, but displace-

[8] Hart, V. L: Congenital dislocation of the hip: early recognition and treatment during first six months of life, Minnesota Med. 32 749, 1949

ment of the femoral head outward and upward to a minimal degree, constituting a subluxation, is more difficult to recognize. Yet it is most important to diagnose a subluxation at this early date, for it is then that replacement results in an excellent anatomic and functional result.

The Newborn. With the infant lying recumbent and both lower extremities held parallel, an anteroposterior view of the hips is taken while traction is exerted on the suspected limb. The procedure is repeated while the extremity is pushed proximally. This "push-pull" film may demonstrate instability of the femoral head. Slight displacements may be detected by the following procedure. A transverse line is drawn on the film through the clear area of acetabulum which represents the triradiate cartilage. From this a perpendicular is erected which passes through the edge of the acetabular roof. Four quadrants are formed where the 2 lines cross. By following the ossified femoral neck upward, the approximate location of the cartilaginous head is ascertained. This head should lie in the inferomedial quadrant. Displacement into the outer inferior quadrant constitutes subluxation, and a position in the outer upper quadrant reveals a frank dislocation.

Figure 61 shows the characteristic findings:

1. Femoral head in upper outer quadrant.
2. Neck appears foreshortened (in reality is anteverted).
3. Acetabulofemoral space is widened.
4. Underborder of femoral neck lies above Shenton's line, whereas normally it is continuous with the upper border of the obturator foramen.
5. The acetabulum is shallow.
6. The acetabular roof is oblique, almost vertical.

Ossification of the femoral head is delayed on the abnormal side Next, the infant's hips are flexed, and the thighs are abducted (Frog position), and the next exposure is made. This reveals the lateral view of the femoral neck which normally forms an angle of 20° to 40° with the shaft. The angulation forward may be increased to as much as 90°, an excessive amount of anteversion. The neck may be shorter than on the opposite side.

The Child. Here the head is ossified, and the findings are more definite. The widening of the joint space, the change of position in push-pull films, the obliquity of the roof, change in contour of the head, and anteversion are easily ascertained. The following sections pertain to refinements in roentgenographic diagnosis which are not absolutely necessary to diagnosis.

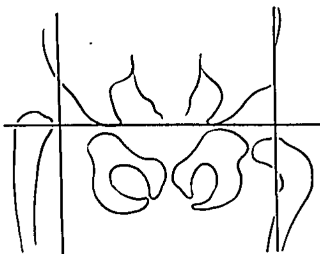


Fig. 61. Identification of dislocation before femoral head ossification. The approximate position of the head is above and medial to the ossified neck. A transverse line is drawn through the top of both triradiate cartilages. This intersects a line dropped from the edge of the acetabular roof, forming 4 quadrants. Normally, the head lies in the lower inner quadrant. The displaced head lies in either of the outer quadrants.

Arthrography. A radiopaque solution such as 17.5 per cent Diodrast or 30 per cent Tenebryl is injected into the joint. The needle is usually inserted anteriorly at a point below Poupart's ligament and lateral to the femoral artery. Information is obtained e.g., the contour and the position of the cartilaginous head, the depth of the acetabulum, and the extent and the constriction of the capsule. The procedure seems to be superfluous inasmuch as failure to obtain reduction by a closed method leads to operative exposure at which time all pathology can be observed and dealt with. The radiopaque solution is completely absorbed within the hour.

CE Angle of Wiberg.[9] This is used after

[9] Wiberg, Gunnar: Studies on dysplastic acetabular and congenital subluxation of the hip joint, Acta chir. scandinav. 83, Suppl. 58, 1939.

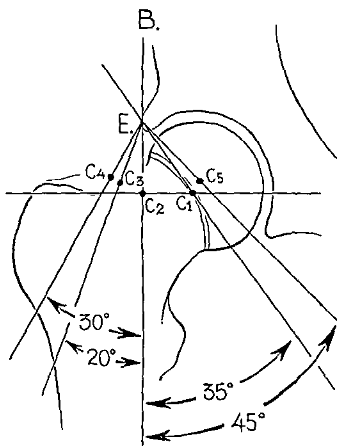


Fig. 62. The CE angle of Wiberg. Variations in CE angle when center of rotation of head is altered. C_1-normal; C_2, C_3, C_4—varying degrees of subluxation; C_2-coxa magna; C_5-coxa plana (center displaced upward). This is used after 3 to 4 years of age when the head is fully ossified. It expresses by a single figure the femoral head and the acetabular relationship. The normal femoral head subtends a perfect circle, and the central point of this circle (C) is the center of rotation which maintains a constant relationship with any point on the acetabulum regardless of the position of the femoral head. The edge of the superior acetabular roof is designated as E. The angle formed by a line passing through this edge from the center of rotation and intersecting the vertical line dropped from this edge is the CE angle. The average normal CE angle is 36°. The normal range varies from 20° to 46°. The center of rotation of the head lies at a point equidistant from the two ends of the epiphyseal line.

the age of 3 to 4 years when the femoral head is fully ossified and its relationship to the *acetabulum is fully established. By this* method, the femoral head-acetabular relationship can be expressed by a single figure.

In the illustration, note the following points: Since the normal head subtends a perfect circle, the central point of the circle denotes the center of rotation of the head, which maintains a constant relationship with any point on the acetabulum, irrespective of the *position of the femoral head. The edge of the* superior acetabular roof is designated as E. The angle formed by a line passing through this edge (E) and the center of the femoral head and intersecting a vertical line (B) dropped from the edge of the roof is the CE angle. The average CE angle is 36°, the normal range varying from 20° to 46°. The center of rotation of the head lies at a point equidistant from the 2 poles of the epiphyseal line and just proximal to the metaphyseal border (C1) The displacement of this center of rotation causes the following variations in the CE angle.

C1: normal

C2, C3, C4: varying degrees of subluxation

C2: coxa magna

C5: coxa plana (center displaced inward)

Acetabular Index. A horizontal line is drawn through the triradiate cartilage. Another is drawn along the roof of the acetabulum and intersects the horizontal line, forming an angle, the acetabular index. This indicates the obliquity of the roof. The normal in the newborn is 27 5°, and in children at 2 years of age it is 20°. As the angle approaches 30°, instability *of the head becomes manifest.*

Determination of the Amount of Anteversion of the Femoral Neck.[10] Generally, it is necessary only to rotate the femur internally during closed or open reduction until the head is firmly fixed within the socket and the full extent of the femoral neck is revealed in outline. The amount of internal rotation of the extremity denotes the degree of anteversion and the necessary *derotation* when corrective osteotomy is done. To determine the amount of anterior torsion accurately the patient is placed supine on the x-ray table, and the hip

[10] Dunlap, K, *et al* · J. Bone & Joint Surg. 35A 289, 1953

is flexed 90°. The thigh is abducted 10°, and the x-ray tube is directed over the hip so that the central ray is perpendicular to the plane of the film. A radiopaque bar is placed lateral to the great trochanter at right angles to the transcondylar axis or frontal plane of the femur. The lateral roentgenogram of the hip thus obtained will exhibit this bar as a reference line which is at right angles to the shaft. A line is drawn through the central axis of the neck of the femur and is projected to meet the shadow of the reference bar. At this point a perpendicular is erected to the reference line. The amount of torsion is the angle formed by this perpendicular line and the line of the central axis of the neck. The normal amount of anteversion is 30° in the infant, which is gradually reduced to 8° in the adult. In congenital dislocation of the hip, the anteversion may increase to 85° or more.

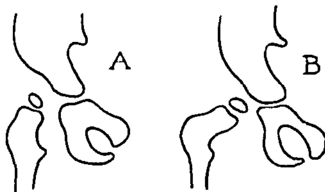


Fig. 63. Anteversion of femoral neck. (A) The limb is in neutral rotation. The neck points forward and appears foreshortened. The epiphyseal ossification center appears small and rounded. The lesser trochanter is prominent. (B) The extremity is rotated internally. The neck in full profile is longer. The ossification center is ovoid and larger. The lesser trochanter is rotated backward and is barely visible.

Treatment

Like other congenital deformities, the earlier treatment is instituted the more likely is a successful result to be realized. Within the first few months, redevelopment of the acetabulum is the normal response of pressure of the femoral head within it. The parts are still mainly cartilaginous and plastic and are easily molded by restoring normal anatomic and physiologic conditions.

During the First Year. Most commonly, only a moderate incomplete displacement or subluxation is found. Merely abducting the limb causes the femoral head to descend toward the center of the acetabulum where

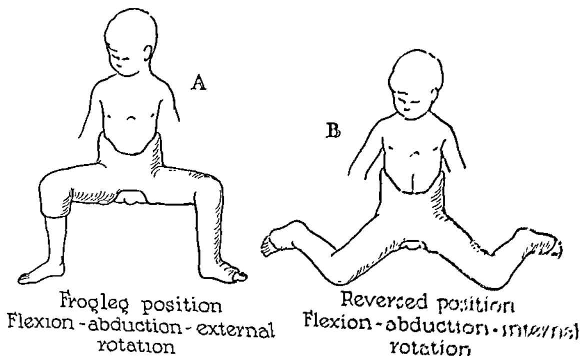


Fig. 64. Retentive casts after reduction of the congenitally dislocated hip.



Fig. 65. Old subluxation of the hip. Originally, this was a complete dislocation which was adequately reduced during infancy, but sufficient deepening of the acetabulum did not take place. Instability has caused erosion of the acetabular roof and degenerative changes throughout the joint. This demonstrates the necessity for prolonged immobilization after reduction of the congenitally dislocated hip.

pressure forces cause the socket to deepen and a roof to form. Whether the condition is unilateral or bilateral, both hips are maintained in abduction continuously for a number of months This can be accomplished by placing a pillow or a specially constructed triangular mattress between the thighs An extra long Denis-Browne or Fillauer splint to which are fixed prewalker shoes can serve the same purpose. Only when roentgenograms demonstrate that the femoral epiphysis lies well within the acetabulum in the inferomedial quadrant and an adequate roof has formed can the splinting be discontinued. This may require anywhere from 3 months to a year.

When a complete dislocation is present, it is usual to find an acetabulum of adequate depth. Reduction is accomplished as follows The infant is recumbent on the table, and an assistant holds the pelvis and the opposite extremity firmly fixed. The dislocated hip is flexed to relax the capsule and adducted to relieve the pull of the adductors. Then the hip is fully flexed. This brings the femoral head around the acetabulum to its shallow postero-inferior aspect. At this point, while the thigh is abducted, the other hand holds the trochanter posteriorly and pushes and guides the head into the socket. Usually, an audible snap or a palpable click is observed as the head passes over the acetabular rim. After reduction, the hamstrings are taut and prevent the knee from being fully extended. One can test stability of reduction by push-pull maneuvers, and roentgenograms confirm the reduction. Bilateral dislocations can be reduced at the same sitting. If the reduction is stable, it may be necessary only to maintain the limbs in wide abduction for several months. However, it is safer to immobilize both extremities in a plaster cast with both hips flexed and abducted, the so-called "frog-leg" position. Both patellae face outward. If anteversion of the neck is suspected, the thighs should be rotated internally so that the patellae face medially. Immobilization is maintained for 3 months, and another cast is applied with the abduction and the rotation partially reduced. Subsequent casts gradually obtain the fully extended and neutral position of the limbs. Manipulation should be done gently to avoid traumatizing the hip. Forceful handling and immediately fully extending the hip (which stretches the capsule and wrings out the blood vessels) has been blamed for subsequent development of degenerative changes in the femoral head which suggests aseptic necrosis (coxa plana). The length of time of immobilization and changes of casts are left to the discretion of the individual operator.

From 1 to 3 Years. Closed reduction is less likely to succeed but always should be attempted inasmuch as a normal hip is obtained frequently enough to warrant conservative measures. A dislocated hip adequately immobilized after an accurate reduction has a good chance to develop normally. The complete dislocation is reduced as described above and retained in plaster for 3 months. During this time, the depth and the rim of the acetabulum should be studied Normally, 3 small ossification centers—the ossa acetabuli anterior, superior, and posterior—appear in the developing rim after the age of 11. However, after the reduction of a dislocation, even at 2 to 3 years of age, the ossification center appears at the upper acetabular rim. This is probably an

attempt at reconstitution of the roof. The appearance of these centers after a reduced dislocation is of value in giving a good prognosis for the acetabulum.[11] Plaster immobilization is continued until the hip is normally reconstituted. An open reduction must be done if closed reduction fails or a tendency to subluxation is noted. Before attempting a closed reduction, it is necessary to apply traction to overcome shortening of the muscles and the fascia sufficiently to allow the head to descend to the level of the acetabulum. The same is a necessary prerequisite to open reduction. When subluxation is present at this age, redevelopment of the acetabulum is inadequate. The acetabulum is ossified and no longer soft and yielding to pressure of the head. The instability and the constant slipping inevitably lead to degenerative arthritis. Surgical deepening of the socket and reconstruction of the roof is necessary.

After the Age of 3. The lack of function leads to shallowness of the acetabulum. The cavity is filled with fibrofatty tissue and adherent capsule. The shortened muscles and fascia keep the femoral head riding high. Marked anteversion is very probable. Attempting reduction at this stage is very traumatizing, and its success is highly improbable. A high percentage of painful stiff hips or redislocations result from nonsurgical treatment. This is especially true in later childhood. After the age of 8, the shortening is too severe to allow reduction of the head into the original acetabulum even by surgical means. Any attempt to restore the head to the normal position should be done before this time. In general, surgical correction is the treatment of choice above the age of 3 but may be undertaken before this time if closed reduction fails.

Surgical Procedures. Selection of the appropriate operation depends on the pathology which is present.

PRELIMINARY TRACTION. The soft tissue structures strongly resist the reduction of the femoral head to the level of the acetabulum. This varies with different individuals and increases with age. A hip which is reduced with the soft tissues tense will be subjected to enormous pressure on the articular surfaces



FIG. 66. Preliminary traction. Bryant traction is sufficient in this case. A more resistant hip requires skeletal balanced traction.

with resultant degenerative changes and possible recurrence of dislocation. Reduction is facilitated if traction is applied preliminary to operation for several days to weeks. Roentgenograms will reveal the descent of the head to the acetabular level. Sometimes it is necessary to perform adductor tenotomies, and even a preliminary operation by which the capsule with the gluteal muscles is stripped from the ilium before skeletal traction is applied. The simpler methods are first tried. Bryant's traction for the younger child and sliding skin traction with a Thomas splint are used for the older child. Adequate descent of the head is absolutely necessary before operating.

OPEN REDUCTION. The anterior iliofemoral or Smith-Petersen incision is used. This skirts along the anterior half of the iliac crest then extends distally between the sartorius and the tensor fascia femoris. The lateral femoral cutaneous nerve is avoided. The gluteus medius and minimus are elevated subperiosteally from the ilium, and the dissection is continued deeply between sartorius and tensor. The reflected head of the rectus femoris above the acetabulum is divided, and the large, elongated, sometimes constricted, capsule is exposed and freed where it is adherent to the

[11] Wiberg, G. Shelf operation in dislocation of the hip, J Bone & Joint Surg 35A:65, 1953.

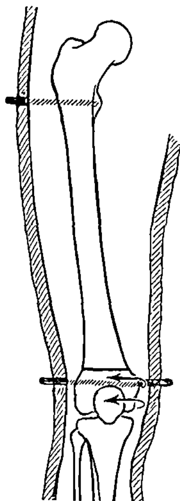


FIG. 67. Operation to correct anteversion of the femoral neck. The entire extremity is rotated internally so that the femoral neck lies in the proper plane. The degree of internal rotation is determined by preoperative x-ray study. A threaded pin inserted into the subtrochanteric area of the femur maintains this position. A threaded pin is inserted into the condylar area of the femur. A supracondylar osteotomy is performed, and the distal fragment is rotated externally until the knee faces forward. The pins are incorporated into the cast.



FIG. 68. Second cast after supracondylar osteotomy to correct anteversion. Distal fragment and leg have been rotated clockwise 90° from the internal rotated position.

ilium, and the upper edge of the acetabulum is encountered. Here the capsule is incised transversely near and parallel with the rim, and the femoral head is exposed. The acetabulum is cleared of the fibrocartilage of the thickened labrum and the fibrofatty haversian gland as well as adherent capsule until the normal articular cartilage is exposed. The depth of the socket can now be ascertained. Some surgeons practice gouging out the car-

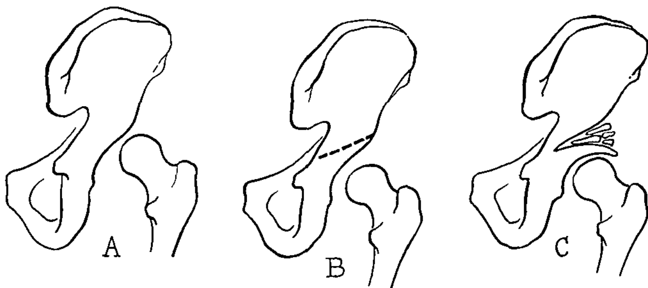


FIG 69. The shelving operation.

tilage to deepen the socket, and they quote excellent results, but degenerative changes probably would seem to be invited by this procedure. While the acetabulum is mainly cartilaginous, ordinary pressure of the reduced head will effect adequate restoration of the socket.

Beyond the age of 5 when bony development of the acetabulum has progressed so that little remodeling can be anticipated, reconstruction of the roof by a shelving procedure can be done at this time. The femoral head is reduced, and the stability is noted. The best position of abduction and the amount of internal rotation required to secure apposition is ascertained and held until the operation is completed and the cast applied. The degree of internal rotation denotes the amount of anteversion to be corrected by osteotomy at a subsequent procedure in 3 or 4 weeks. The ligamentum teres is excised. It may be necessary to elongate the rectus femoris and the sartorius before the head can be adequately replaced. The capsule is also partially excised and shortened and sewed firmly to aid in stabilizing the head. Postoperatively, the cast extends from the nipples to the toes and to the thigh on the opposite side.

Correction of Anteversion. Inasmuch as anteversion increases with age and persistence of the abnormal position of the head, correction is indicated more frequently after the age of 3. A section of the cast is removed from the lower third of the thigh, and the lateral aspect of the femur is exposed. A threaded pin is drilled into the bone and extends out of the operative wound to be held in the plaster for fixation of the upper fragment. While the pin is held firmly, the bone is osteotomized, and the distal fragment is rotated until the patella

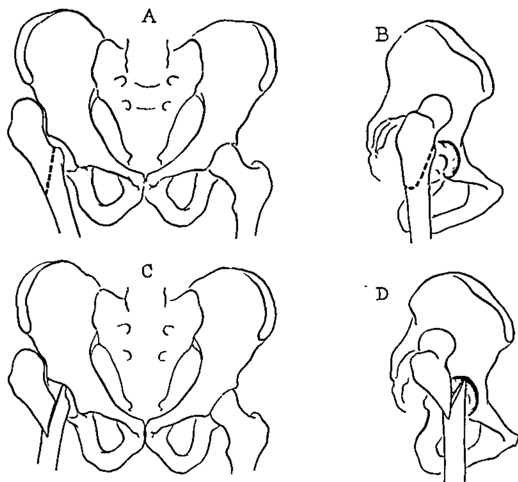


Fig. 70. Lorenz bifurcation osteotomy. (A and B) Direction and level of section. (C and D) Position after displacement of distal fragment into the acetabulum.

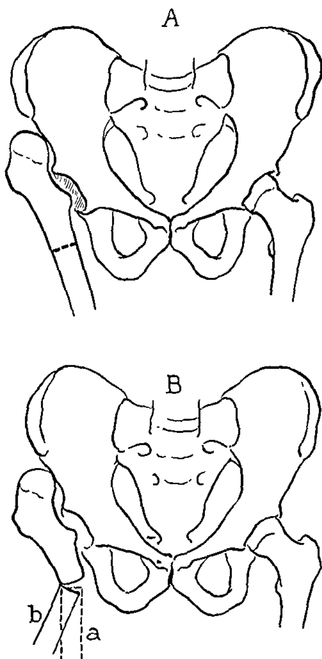


FIG. 71 Shanz osteotomy. (A) Preoperatively. A film taken with the limb completely adducted will indicate the angle of correction The level of section is well shown. (B) Position after osteotomy. The usually accepted positioning is at *a*; *b* hyperabduction indicated for extreme shortening and instability.

points forward. After closure of the wound, the two sections of the cast are joined. The pin is removed when union is observed on roentgenograms.

SHELF OPERATION. The older the patient when reduction is effected, the less likely is the deepening of the acetabulum, the greater the subsequent disability and inevitable symptoms of pain, stiffness and fatigue. The roof can be reformed at the same time as the reduction. However, it is best to do it after the age of 5 when the ossification within and about the acetabulum is sufficient to ensure success of this bone plastic procedure. Preliminary traction for several weeks to overcome the muscle pull is necessary to lessen the danger of the femoral head's being pulled proximally against the newly formed buttress and compromising the result. At operation all soft tissue structures which may similarly interfere are dealt with. Then a cortical flap of bone, starting immediately above the upper acetabular rim and extending inward and downward, skirting the articular aspect, is turned downward over the head and is maintained by insertion of bone blocks and chips. Traction must be continued postoperatively until bony consolidation is complete.

In irreducible dislocations, a shelf operation is necessary when instability causes symptoms The lumbar lordosis resulting from posterior position of the femur may cause low back pain. At operation, the femoral head is brought forward to a point just above the acetabulum, and the shelf is constructed. The pelvic tilt and lordosis are reduced, and stability is effected. This procedure is likely to be successful when performed after the age of 10.

STABILIZING OSTEOTOMIES. At a late age, stabilizing osteotomies are the procedures of choice to eliminate pain at the site of the false acetabulum on the wing of the ilium. A bony support on the femur is formed. Two main types are used—the Lorenz and the Schanz.

Lorenz Bifurcation Osteotomy. A long oblique osteotomy in the subtrochanteric area is made so that the proximal end of the distal fragment lies at the level of the acetabulum. The plane of the osteotome is directed from the distal end at the posterolateral aspect toward the proximal end at the anteromedial aspect of the bone. Then the limb is abducted and extended so that the proximal end of the distal fragment is directed medially and anteriorly into the acetabulum Such large raw surfaces of the osteotomized bones are apposed to each other that union in the angulated position takes place readily. A cast is applied for about 3 months or until union is demonstrated by x-ray examination.

Schanz Osteotomy. By this procedure the femur is sectioned transversely at the lower border of the pelvis, and the upper fragment is angled inward until it rests against the side wall of the pelvis. Thus, the body weight gains a bony support, and stability is demonstrated by improvement of the Trendelenburg sign. The adduction of the proximal fragment lengthens the distance of the gluteus medius and provides a fulcrum so that adequate leverage of the muscle is obtained. Preliminary to operation, an anteroposterior roentgenogram is taken with the lower extremity completely adducted. A measuring stick with radiopaque markers is placed alongside the limb, and the level of osteotomy and the degree of angulation are ascertained. A Vitallium plate is prepared and angulated sufficiently not only to deviate the upper fragment medially against the pelvic wall but also anteriorly to overcome the posterior displacement of the upper femur. At operation the bone is sectioned transversely, and the plate is attached to the upper fragment. Then the distal shaft is abducted and extended and approximated to the distal half of the plate which is then attached. The cast is generally necessary for about 3 months. This procedure is contraindicated before the age of 15 because the loss of angulation is common during the growth period.

CONGENITAL CLUBFOOT

Definition

Congenital clubfoot (talipes) is a gross deformity of the foot present at birth. The direction of deformity of the individual components of the foot is described by the following terms:

Equinus. The forefoot is plantar flexed (at ankle and midtarsal areas).

Calcaneus. The forefoot is dorsal flexed. The calcaneus forms the plantar prominence.

Varus. Heel and forefoot are inverted. The plantar surface faces medially.

Valgus. Heel and forefoot are everted. The plantar surface faces laterally.

For example, talipes equinovarus, the most common type (95%), describes the foot which is plantar flexed and inverted. Talipes calcaneovalgus, calcaneovarus and equinovalgus are the other less common types.

Etiology

The causative factor is unknown. The theories are:

1. *Intra-uterine compression.*
2. *Arrest in fetal development.* This would explain frequently associated anomalies.
3. *Dysplasia of muscles*[12] causes muscle imbalance.
4. *Abnormal tendon insertion,* particularly anterior tibial.

Deformities of feet which are due to obvious causes are not usually included in this discussion of clubfoot, because the exciting cause demands primary consideration in treatment. These conditions which will be discussed under the appropriate headings include cen-

[12] Garceau, G. J.: Talipes equinovarus, Am. Acad. Orthop. Surg. Instructional Course Lectures 7:119, 1950.

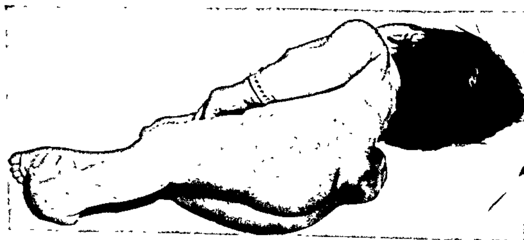


Fig. 72. Spina bifida. The associated club foot deformity is severe.



FIG. 73. Talipes equinovarus deformity. Note the internal tibial torsion.

tral nervous system diseases (spina bifida, poliomyelitis, Friedrich's ataxia), imperfect muscle development (arthrogryposis), lymphatic stasis (congenital constriction band) and absent bony structures (tibia, etc.).

Brief mention should be made of spina bifida, which is not always clinically evident, but roentgenographic investigation for it always should be conducted in cases of clubfoot deformity. When the spinal defect is large and associated with extrusion of spinal contents, muscle imbalance, sphincter paralysis and sensory loss accompany a severe foot deformity.

Pathology

In talipes equinovarus the leg muscles are smaller than in the normal leg and microscopically may show signs of possible degeneration. The tendo achillis passes downward and medially toward the calcaneus, thereby helping to maintain the bone in inversion. The ligaments between the calcaneus, the talus and the navicular are thickened and contracted. The navicular is displaced downward and medially and is maintained by the shortened posterior tibial tendon and the contracted ligaments, including the deltoid ligament. The anterior end of the os calcis is displaced medially beneath the head of the astragalus. The plantar aponeurosis is thickened and contracted along with the short plantar muscles so that a high arch deformity (cavus) is created, and the forefoot is held in equinus The forefoot is adducted and inverted by the combined action of the tight anterior and posterior tibial muscles.

At first the bony structures are normal. Later they become deformed as an adaptation to their persistently abnormal position. The top of the talus becomes flattened. The talar head enlarges. The calcaneus angulates medially. All metatarsals are curved medially. The cuneiforms and the cuboid become wedge-shaped with their bases disposed dorsolaterally. Internal torsion of the tibia is a frequently associated deformity.

In talipes calcaneovalgus, which constitutes the major part of the remaining clubfoot types, no definite pathology can be ascertained, because the deformity usually corrects spontaneously or with a minimum of treatment. However, should the deformity persist, the scaphoid displaces laterally with the forefoot structures, the head of the talus points medially and inferiorly, and the longitudinal arch is flattened. All ligamentous structures are lax and elongated.

Clinical Findings

The deformity is divisible into 3 main components:

1. *Equinus*, in which the forefoot is dropped plantarward.

A. *At the ankle*—heel cord tight

B. *At midtarsal area*—plantar structures tight

2. *Varus of heel.* The tight medially inserted heelcord and medially contracted ligaments resist correction.

3 *Adduction and varus of forefoot.* The anterior and posterior tibial muscles pull the first metatarsal and the scaphoid into inversion.

In addition, the contracted plantar aponeurosis and muscles create a cavus deformity. The anterior end of the talus forms a dorsal and lateral bony prominence The tibia is twisted inwardly. The deficient musculature causes an atrophic appearance of the leg. Flexibility of the foot is lost to a varying degree, depending on the severity and the age of the deformity.

Roentgenographic Findings

Normally, the talus, the scaphoid, the inner cuneiform and the first metatarsal form a straight line. In a clubfoot the scaphoid is displaced medially and inferiorly to the head of the talus, carrying the cuneiform and the



Fig. 74. Manipulation of talipes equinovarus. The tips of the thumbs form the fulcrum against which the fingers of the left hand evert the heel and the fingers of the right hand abduct and evert the metatarsals. Pressure is gentle, maintained a few seconds and repeated.

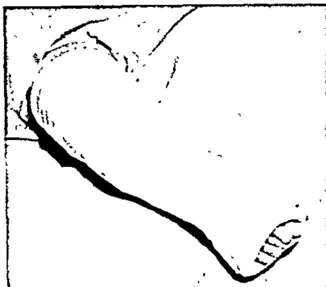
metatarsal with it. The center of ossification for the scaphoid normally does not appear before the third or the fourth year, but the position of the cuneiform and the metatarsal indicates displacement of the scaphoid. The shadows of the talus and the calcaneus normally overlap except at the anterior end where the calcaneus is displaced laterally so that the axis of the calcaneus is in line with the 4th and the 5th metatarsals. In talipes equinovarus the anterior end of the os calcis is displaced medially and overlaps the talar head. Correction demands that the medial and lateral axes be restored.

When the deformity has persisted for several years, the metatarsals become curved medially, the scaphoid becomes deformed and enlarged medially while the talar head is asymmetrical and enlarged laterally and forms an obstacle to reduction. An anteroposterior view of the os calcis reveals a varus deformity of the bone. The body of the talus is in equinus, only its posterior portion articulates with the tibia and is flattened and sclerotic.

In calcaneovalgus, x-ray films are normal at first, but as time passes the scaphoid comes to lie lateral to the talar head, which in turn presents medially and plantarward. A lateral weight-bearing view reveals flattening of the longitudinal arch, evidence of ligamentous inadequacy.

Diagnosis

Assuming that no primary causative factors can be discovered, the deformity must be regarded as the typical primarily occurring clubfoot. The condition must be differentiated



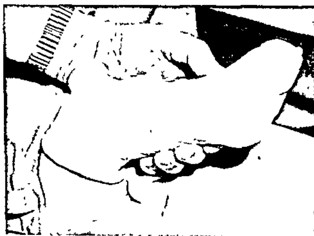


Fig. 75. The finished cast. Note the equinus position during correction of the adduction and the varus. The cast extends above the knee, which is in the flexed position to prevent rotation of the limb within the cast, and relaxes the calf muscles.



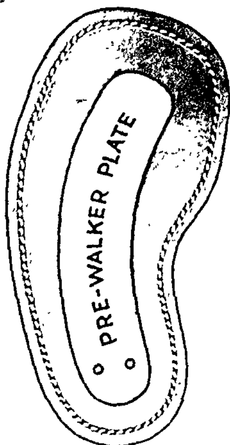


Fig. 76. Prewalker clubfoot shoe. This is used to *maintain* correction obtained by other means. It has no corrective function. Note the pronounced outflare of the forepart of the shoe, the retentive strap and buckle across the ankle, and the flat steel plate over the sole. (I. Sabel Company, Philadelphia)

from the "Z" foot. which consists of metatarsus adductus and a relaxed valgus of the rear foot. This is a special entity in which the scaphoid lies lateral to the talus so that treatment by the usual method will displace the scaphoid further and produce a severe flatfoot.

Treatment

Conservative treatment consists of gradual manipulative reduction of the deformity and maintenance by retentive apparatus. This treatment is applicable to cases seen early, within the first year or two.

Surgical intervention is reserved for the clubfoot which resists conservative treatment or is seen at a late date after the deformity has become fixed by the changing contour of the bony structures. The longer conservative treatment is delayed, the more resistant will be the deformity, and the greater the need for surgical correction.

Conservative Treatment. Manipulation is directed to stretching the contracted tissues gradually and repeatedly over a period of time —weeks to months No anesthesia is necessary. Gentleness and deliberateness is the essence of treatment. The adduction and varus components are corrected first. The foot is grasped by both hands so that the thumb tips press over the lateral bony prominence formed by the cuboid and the base of the first metatarsal. This acts as the fulcrum while fingers about the heel and the metatarsals pull these structures into abduction and eversion. This is done gently and repeatedly. Forceful manipulation tears the contracted tissue medially, resulting in cicatrix; the joints will be damaged and eventually will become ankylosed. A stiff permanently deformed foot would result.

When the deformity can be partially corrected with a minimum of pressure, a cast is applied, maintaining a position just short of the obtained correction. An assistant holds the limb with the knee flexed to a right angle while another assistant holds the foot in the corrected position. A few layers of protective sheet wadding are applied, and the cast bandage is rolled on smoothly from toes to upper thigh. While the plaster is setting, the surgeon holds the foot in the corrected position. Attempts to obtain correction after applying the plaster will cause points of pressure necrosis and constriction of the foot and the leg. The mother is alerted to watch for danger signs

over the ensuing hours. Cyanosis and swelling of the toes and continued irritability and crying by the infant are sufficient indication for removal of the cast and relief of pressure. At 2-week intervals the cast is removed, and the procedure is repeated, a little more correction being secured at each visit. When varus and adduction have been overcome (as denoted by x-ray realignment of the axes), correction of the equinus is initiated. If this is started while the scaphoid is still displaced medially, recurrence of the deformity is the rule, because with weight-bearing the talus will push on one side of the scaphoid and redisplace it. The foot should not be brought up before proper axial alignment has been obtained. Manipulation by stretching the calf muscles and the posterior capsule of the ankle is accomplished by direct upward pressure on the anterior end of the os calcis. Pressure distal to the midtarsal joints causes the small tarsal bones to move dorsalward, resulting in a "rocker-bottom" foot. Repeated stretchings and castings are done until adequate dorsiflexion has been obtained.

The tendency to recurrence is strong. Therefore, overcorrection to some degree toward the opposite deformity is advised by some authorities. However, Kite[13] states that overcorrection of the scaphoid displacement leads to another disabling deformity, a flatfoot in which the scaphoid lies lateral and dorsal while the talar head protrudes medially and plantarward. Nevertheless, persistence is necessary until complete anatomic restoration is attained. The correction is maintained by daily, continued, repeated manipulations and stretchings by the parents. The infant wears a "prewalker" shoe to aid in holding the correction until he can stand and walk. This is a high-top shoe with an outflare last, a steel sole plate, a rigid extended counter, and a strap across the anterior aspect of the ankle to hold the foot snugly in the shoe.

The Denis-Brown splint is also used to preserve correction. This consists of a metal bar with a sole plate at either end to which the feet are attached by adhesive bandaging. On the treated side, the plate is bent outward so that the foot is kept tilted in eversion, abduction and external rotation. Theoretically, the child's kicking motions aid the correction. The Fillauer splint is a modification using clamps to hold shoes at the outer ends of the splint, which simplifies its application. The mother need only to insert the infant's feet in the shoes which are already fixed in the appropriate position. The method does not eliminate the need for manipulations, particularly heel-cord stretchings. By maintaining the feet continuously in external rotation, the internal tibial torsion may gradually be overcome.

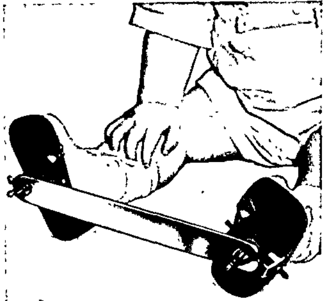


FIG. 77. Fillauer splint. This removable splint is useful for treating clubfoot and rotational deformities of the lower extremity. The shoe clamps may be rotated internally and externally. The flat intervening bar may be bent to hold the feet in inversion or eversion. An oversized bar will maintain wide abduction useful in treating dysplasia of the hip or after reduction of a dislocated hip. (The Fillauer Company, Chattanooga, Tenn.)

Recurrence of deformity demands return to the original cast procedure. Resistant deformity should be overcome surgically.

Surgical Treatment. Operations are confined to the soft tissues when done before the age of 8 or 9. Before this time the bony structures are not sufficiently ossified for fusion so that recurrence is inevitable.

SOFT TISSUE OPERATION.[14] This is based on

[13] Kite, J H · Am Acad Orth. Surg, Instructional Course Lectures 7:116, 1950.

[14] Brockman, E P . Congenital Clubfoot, London, Wright, 1930.

Brockman's premise that a congenital atresia of the socket for the head of the talus is the basis for the deformity. The shortened muscles acting upon the scaphoid must be lengthened and the socket enlarged.

First, through a lateral incision, the plantar aponeurosis and muscles are detached from the os calcis Through a medial incision the severance is completed. The deltoid ligament is cut free from the medial malleolus. The poste-

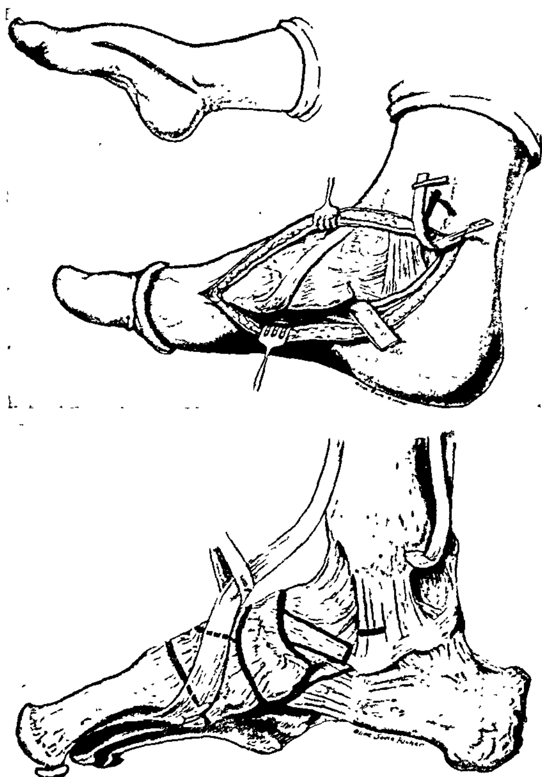


Fig. 78 Soft tissue operation for clubfoot. (A, *top*) Incision. Section of posterior tibial tendon (B, *bottom*) Depicting normal ligamentous structures. Lines indicate ligaments to be excised. (*Continued on facing page*)

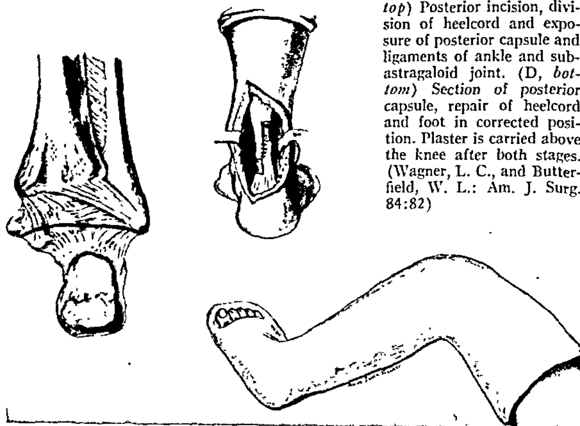


Fig. 78. (*Cont.*) Soft tissue operation for clubfoot. (C, *top*) Posterior incision, division of heelcord and exposure of posterior capsule and ligaments of ankle and subastragaloid joint. (D, *bottom*) Section of posterior capsule, repair of heelcord and foot in corrected position. Plaster is carried above the knee after both stages. (Wagner, L. C., and Butterfield, W. L.: Am. J. Surg. 84:82)

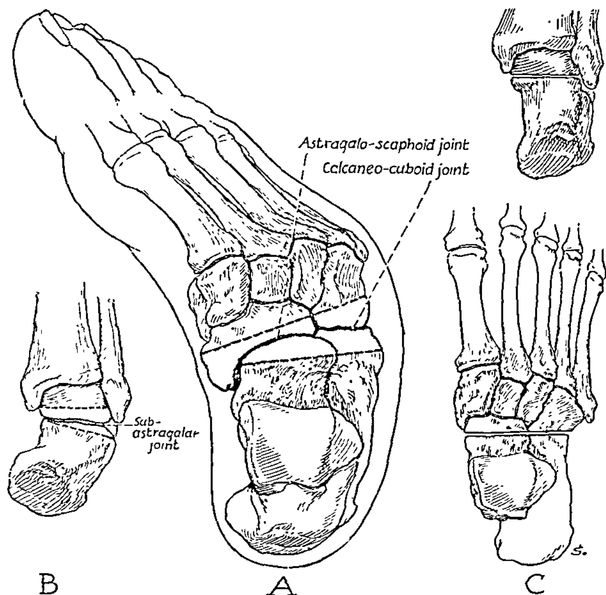


FIG. 79. Bony reconstruction for neglected equinovarus deformity. The wedges are removed from the midtarsal and the astragalocalcaneal joints with their bases directed toward the direction of convexity. (Speed, J. S., and Knight, R. A.: Campbell's Operative Orthopaedics, St. Louis, Mosby)

rior tibial tendon is cut within its sheath. The inferior aspect of the tarsal bones is cleared of all ligamentous structures, cutting particularly the spring ligament which connects the articulation between the calcaneus and the scaphoid. Abduction and dorsiflexion should easily replace the scaphoid in front of the talus and move the anterior end of the calcaneus outward. A cast is applied with only partial correction until the operative wound is healed, and then the foot correction is completed. Later, an Achilles tendon lengthening may be required to correct the equinus. Finally physical therapy directed to strengthening the dorsiflexor and evertor muscles completes and maintains the correction. When weakness of the peroneals is demonstrable, the anterior tibial may be transferred to a new insertion at the lateral side of the foot.

BONE OPERATION. Severe or old deformities require resection of bone for correction, and fusion for maintaining the correction. Essentially, it consists of removal of a wedge of bone with the base directed laterally and dorsally to overcome varus and adduction. First, the attachments of the plantar aponeurosis and short flexor muscles are stripped from the os calcis. A bony wedge is removed at the level of the calcaneocuboid and astragaloscaphoid joints, the base being lateral. This corrects the adduction. Another wedge is removed from the subastragalar joint, again

with the base lateral. This corrects varus. Achilles tendon lengthening completes the procedure. The corrected position should then be secured with ease and a cast is applied to be worn for three months. Subsequently, a foot and ankle brace or leather corset guards against recurrence until fusion is solid.

An alternate bone procedure can be done when the deformity is severe in a child too young for fusion.[15] Through small puncture wounds in the cuboid and the anterior portion of the talus and the os calcis, the cancellous bone is curetted out. The cortical exterior of these bones remains intact but is pliable and yields to bending force. Correction with a Thomas wrench is done. Occasionally, removal of the lateral cortex of the os calcis and the cuboid may be done to permit collapse. The articulating surfaces are undisturbed. Preliminary Achilles tendon lengthening and plantar stripping should be done.

Calcaneovalgus Foot. The cause of this deformity is suggested as positional compression in utero, because of the strong tendency to spontaneous correction. The talus is plantar flexed, the navicular is displaced lateralward in relation to the talar head, and the calcaneus is everted. Clinically, the head of the talus forms a medial and plantar bony prominence, and the longitudinal arch is flattened. Marked relaxation of all ligaments is evident. The foot can be passively dorsiflexed to the extreme so that it touches the anterior aspect of the foot. In standing, the child appears to be standing on the medial side of the foot. The forefoot is abducted, and the position is accentuated by the associated external rotation of the tibia The infant, when sleeping in the prone position, usually holds the limb in the abnormal position, thereby perpetuating the deformity. Walking is delayed and slow. Pain due to foot strain and pressure over the medial bony prominence is complained of. Roentgenograms reveal a sag in the midtarsal area, and the talus points downward and medially. At first the scaphoid may lie in its correct position but later may shift laterally as the flatfoot deformity persists.

Treatment consists of manipulating and splinting into the opposite direction, i.e., plantar flexion, adduction and inversion. This can be done by stretching manipulations and casts and followed later with a Fillauer splint holding the feet in marked internal rotation. Maintenance of correction must be stubbornly persisted in until full bony development will resist redisplacement. The foot is stretched daily, and exercises are done to redevelop the anterior and the posterior tibials. Oxford shoes with an inflare last, medial wedges in the soles, and arch supports are worn. The foot with extreme laxity of ligaments is prone to recurrence. Shoe supports may become necessary throughout childhood until corrective arthrodeses, such as described under Flatfoot, can be done. As a matter of fact, these cases may be regarded as congenital flatfeet and treated as such.[16] The parents should be instructed that treatment may fail and a persistent flatfoot may result, but that such a deformity is not necessarily disabling.

METATARSUS ADDUCTOVARUS[17, 18, 19]
(Metatarsus Varus)

This congenital deformity, which appears to be increasing in frequency, consists of medial displacement of the first metatarsal, inversion of the forepart of the foot, and a cavus arch.

At birth, in contrast with clubfoot, the deformity is not evident. However, within the first few weeks the big toe and the first metatarsal are pulled medially by strong active contractions of the abductor hallucis muscle. Next, as the deformity increases, the forepart of the foot is inverted (varus) and pulled upward by the anterior tibial. This muscle has an anomalous insertion into the medial side of the first metatarsal and passes to the inferomedial aspect of the foot. This action eventually elevates the longitudinal arch into a cavus. Eventually, the adducto-cavo-varus deformity becomes fixed, all the metatarsals being drawn medially and inverted. When weight is borne on this foot, in order that the

[15] Curtis, F. E, and Muro, F.: Decancellation of os calcis, astragalus, and cuboid in correction of talipes equinovarus, J. Bone & Joint Surg 16:110, 1934.

[16] Thomson, J. E M: Treatment of congenital flatfoot, J. Bone & Joint Surg 28:787, 1946.

[17] Peabody, C. W., and Muro, F: Congenital metatarsus varus, J. Bone & Joint Surg 15 171, 1933.

[18] Kite, J. H: Congenital metatarsus varus, J. Bone & Joint Surg 32A·500, 1950

[19] McCormick, D. W., and Blount, W P.: Metatarsus adductovarus, J.A.M.A 141:449, 1949.

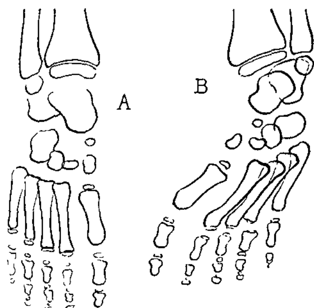


FIG 80. Congenital metatarsus varus (A) The talus, the scaphoid, the first cuneiform and the first metatarsal form a straight line. The anterior ends of the talus and the calcaneus are separated. (B) The first metatarsal is carried medially and is in line only with the inner cuneiform and the scaphoid, the latter lying lateral to the talar head. The talus and the calcaneus are in the flatfoot position, the anterior ends lying in a divergent relationship The inversion of the forefoot causes the cuneiforms to overlap and the lateral aspect of the metatarsals to be visualized. The metatarsals normally are bowed dorsalward and in this view they are wrongly identified as deformed.

medial border of the foot make contact with the floor the foot rotates on its longitudinal axis so that the hindpart of the foot, or calcaneus, is thrown into eversion or valgus. Finally, in the adult, the adducted first metatarsal forms a bunion, and the big toe deviates outward (hallux valgus) in response to pressure forces of wearing a shoe. Thus, what first appears to be a mild deformity, an ordinary intoeing of the large toe, has serious long-range complications. With advancing age the "serpentine foot" develops, consisting of adduction and varus of the metatarsals, hallux valgus, valgus of the calcaneus, and cavus.

Early Clinical Picture

If treatment is to be successful, the condition must be recognized before the deformity is severe and fixed. An excessively acting abductor hallucis can be determined by stimulating or stroking the medial border of the foot, whereupon the big toe bends medially, and the first metatarsal spreads from the second. Normally, during infancy no longitudinal arch is present. By taking an imprint of the sole, early cavus is detected. Normally, the tarsal area is mobile, but in metatarsus adductovarus the distal tarsals are resistant to passive attempts to swing the metatarsals into abduction. Later, this rigidity is so firm that the first metatarsal lies in a permanently adducted position. The heel now lies in valgus. A bony prominence on the lateral border consists of the dorsal aspect of the cuboid and the base of the fifth metatarsal.

Roentgenographic Findings

The findings are characteristic as the deformity develops. The metatarsals are adducted. The base of the first metatarsal articulates with the medial aspect of the first cuneiform, which is irregular in shape. The ossification centers for the first and the second cuneiforms are delayed. The great toe is widely separated from the second toe. Valgus of the heel is evidenced by wide separation of the anterior ends of the calcaneus and the talus. The scaphoid lies lateral to the head of the talus in the flatfoot position. When varus or inversion is present, the metatarsals are rotated about their axes so that their dorsal curves are visible in the A-P view. This produces *an apparent medial curving* of the metatarsals.

Treatment

Early treatment consists of manipulations and retention casts. With the cuboid as a fulcrum, the forepart of the foot is swung into abduction. While the cast is being applied, the heel is held in varus to guard against development of a flatfoot. After the age of 6 months, it may be necessary to overcome the distal tarsal rigidity under anesthesia. Subsequent weekly stretchings and castings are done without anesthesia until overcorrection is obtained. Occasionally, it becomes necessary to sever the abductor hallucis tendon and the metatarsal attachments of the anterior tibial. In the late neglected case, a bone wedge is removed from the distal tarsal row, and the first metatarso-cuneiform joint is arthrodesed in the corrected

position. The Keller procedure—resection of the base of the proximal phalanx of the big toe—corrects the hallux valgus. Tendon operations can be done in infancy and childhood, but bone procedures must await full bone development, usually after the age of 10.

When in infancy correction has been obtained by conservative means, manipulations should be persisted in even after discontinuing the casts. Swungout shoes are worn, but one must guard against pressure being exerted upon the toes alone and development of a hallux valgus deformity.

CONGENITAL HALLUX VARUS

Congenital hallux varus is a deformity found at birth, consisting of medial angulation of the large toe at the metatarsophalangeal joint. One or more of the following conditions are present:[20] (1) the first metatarsal bone is usually short and thick; (2) accessory bone and toes are often associated; (3) varus deformity of one or more of the other metatarsal bones; (4) a firm fibrous band extends from the inner side of the large toe to the base of the first metatarsal. Over the medial aspect of the first metatarsal, an articulation with a small accessory bone represents a vestigial accessory toe. This may be the incompletely developed medial segment of a primary double hallux.

Other causes of hallux varus include: overcorrection of a bunion operation, paralysis of the adductor hallucis, infection about the metatarsophalangeal joint, and malunion of fractures about the joint.

TREATMENT

The correction is surgical. Through a dorsal longitudinal incision, the interspace between the first 2 metatarsal heads is developed, and bone is excised from the lateral aspect of the head of the first metatarsal The capsule is freed from about the dorsal, medial, and plantar aspects of the metatarsal head. Next, the accessory bone, the fibrous band and the medial sesamoid are removed from the medial side of the metatarsal. Then freeing of the capsule from the metatarsal is completed At this stage the toe can easily be displaced laterally. The extensor hallucis brevis tendon is severed at its junction with the muscle, and while the toe is held laterally in the corrected position the tendon is routed through a drill-hole in the metatarsal neck. The lateral capsule is imbricated to maintain the new position. Postoperatively, firm bandaging to the other toes is all that is necessary.

[20] McElvenny, R. T · Hallux varus, Quart. Bull. Northwestern Univ. M School 15 277, 1941.



FIG. 81. Overlapping little toe. The toe is short, dorsiflexed, rotated and adducted.

CONGENITAL OVERLAPPING LITTLE TOE

In this condition, which is usually bilateral, the small toe is dorsiflexed, rotated so that its dorsal surface faces laterally, and is adducted and overlies the 4th toe. By passively flexing the toe, the taut extensor tendon stands out prominently as the factor preventing reduction of the deformity. When the deformity is mild, passive stretching into flexion and abduction may suffice to overcome it. Surgical correction is usually necessary. Severance of the extensor tendon and incising the dorsal capsule of the metatarsophalangeal joint allows the toe to drop down. The extensor tendon reunites in an elongated position.

An alternative procedure consists of cutting the extensor tendon proximally and freeing it to its insertion which is left intact. Then the tendon is routed through the medial aspect of the toe to its plantar surface, then laterally and joined to the tendons of the flexor and the abductor, thus, active movements of these muscles aid in correction. Where the cosmetic factor does not enter into the decision, amputation of the toe provides a normally functioning foot.

CONGENITAL DISLOCATION OF THE KNEE

(Congenital Genu Recurvatum)

One or both knees may be found at birth in a position of hyperextension, and in extreme cases it may be possible to approximate the leg to the anterior surface of the thigh. The proximal end of the tibia is displaced anteriorly and laterally on the femur. The quadriceps tendon and the iliotibial band are shortened The patella is small or absent but begins forming after correction of the deformity. The anterior half of the capsule is contracted, but the posterior portion and the anterior cruciate ligament are elongated and lax.

Two types are recognized. The *traumatic developmental type* is due to malposition in utero or fibrofatty degeneration of the quadriceps. The second type is a *primary embryonic defect* as indicated by the accompanying defects such as harelip and clubfoot. The first type is far more common

Conservative treatment consists of repeated manipulations and castings to permit gradual flexion. Subcutaneous tenotomy of the iliotibial band is often necessary, and braces may be required to maintain reduction.

For severe resistant cases, surgical correction involves lengthening or sectioning the quadriceps tendon, the iliotibial band, the anterior capsule and the posterior cruciate ligament. The posterior capsule and the anterior cruciate ligament are reefed. A maldisplaced gastrocnemius should have its proximal ends anchored to the femoral condyles.[21]

CONGENITAL COXA VARA

(Infantile Coxa Vara; Developmental Coxa Vara)[22, 23, 24]

Normally, the infantile femoral neck forms an angle with the shaft of 120° to 140°. Reduction to a more acute angle constitutes a coxa vara deformity and is due to a variety of causes including congenital, infection, trauma, tumor and slipped upper femoral epiphysis. Congenital coxa vara is that type found at birth or shortly thereafter. Pathologically, it consists of a progressively increasing acuteness of the neck-shaft angle; shortness of the neck; a vertical direction of the epiphyseal plate; an oblique defect in the neck which extends from the proximal medial to the distal lateral borders of the neck and is composed of cartilage and osteoid tissue; a great

[21] Clayburgh, B J.: Congenital dislocation of the knee, Proc. Staff Meet, Mayo Clin 30 396, 1955

[22] Zadek, I: Congenital coxa vara, Arch Surg 30 62, 1935.

[23] LeMesurier, A B: Developmental coxa vara, J Bone & Joint Surg. 30B:595, 1948.

[24] Babb, F S, Ghormley, R. K., and Chatterton, C. C.: Congenital coxa vara, J Bone & Joint Surg 31A.115, 1949.

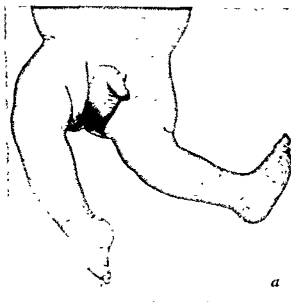




FIG. 82. Congenital dislocation of the knee. (Clayburgh, B. J.: Proc. Staff Meet. Mayo Clin. 30:396)

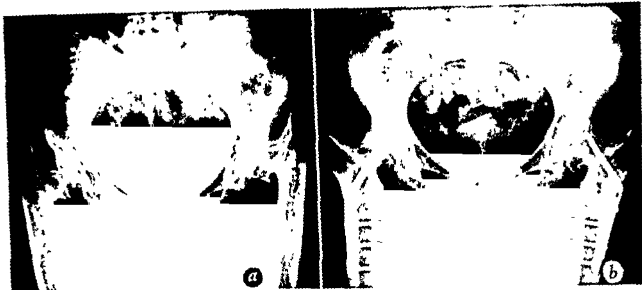


Fig. 83. Congenital bilateral coxa vara. Pain, weakness, and waddling gait. Bilateral Shanz osteotomy done to increase efficiency of gluteus medius muscle and to change the weight-bearing angle and the surfaces of femoral head. Vitallium mold arthroplasty done on left for degenerative changes. (Coventry, M. B.: Proc. Staff Meet. Mayo Clin. 29:48)



trochanter extending upward toward the ilium as a beak; a shortened femur; and secondary degenerative changes in the acetabulum due to malapposition. Histologically, the head of the femur is normal.

Clinical Picture

Clinically, a *painless limp* is the usual complaint. On examination, one finds a *shortened lower extremity* and a *positive Trendelenburg* test, the latter because the shortened distance from origin to insertion has weakened the gluteus medius. *The greater trochanter lies above Nelaton's line.* When the condition is bilateral, the bilaterally weakened hip abductors cause a waddling gait, and the *increased lumbar lordosis* suggests dislocated hips, but no telescoping is demonstrable. The neglected case in adult life has degenerative arthritic symptoms superimposed such as pain, stiffness and weakness. A *progressive limitation of abduction and internal rotation* is noted.

Roentgenographic Findings

Roentgenographic findings are typical. The oblique defect in the femoral neck extending upward toward the proximal portion of the vertically disposed epiphyseal plate forms with the latter an inverted V or Y. This defect has the appearance of a nonunion. The triangular fragment may be irregular and fragmented. The neck is short, and the greater trochanter is beaked. The head is large and somewhat translucent. One unit, consisting of the head, epiphyseal cartilage and the triangular fragment, appears to be slipping downward.

Treatment

Treatment is aimed at (1) *obliterating the neck defect* and thereby halting progression of the deformity, and (2) *correcting the already existing deformity.* A subtrochanteric osteotomy and widely abducting the distal fragment will convert the oblique almost vertical defect, which is perpetuated by shearing stresses, into a horizontally displaced defect, which will fuse or unite by exposure to com-

CONGENITAL DISLOCATION OF THE KNEE

(Congenital Genu Recurvatum)

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Conservative treatment consists of repeated manipulations and castings to permit gradual flexion. Subcutaneous tenotomy of the iliotibial band is often necessary, and braces may be required to maintain reduction.

For severe resistant cases, surgical correction involves lengthening or sectioning the quadriceps tendon, the iliotibial band, the anterior capsule and the posterior cruciate ligament. The posterior capsule and the anterior cruciate ligament are reefed. A maldisplaced gastrocnemius should have its proximal ends anchored to the femoral condyles.[21]

CONGENITAL COXA VARA

(Infantile Coxa Vara; Developmental Coxa Vara)[22, 23, 24]

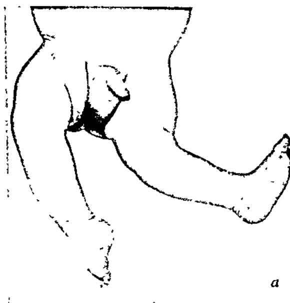
Normally, the infantile femoral neck forms an angle with the shaft of 120° to 140°. Reduction to a more acute angle constitutes a coxa vara deformity and is due to a variety of causes including congenital, infection, trauma, tumor and slipped upper femoral epiphysis. Congenital coxa vara is that type found at birth or shortly thereafter. Pathologically, it consists of a progressively increasing acuteness of the neck-shaft angle; shortness of the neck; a vertical direction of the epiphyseal plate; an oblique defect in the neck which extends from the proximal medial to the distal lateral borders of the neck and is composed of cartilage and osteoid tissue; a great

[21] Clayburgh, B. J.: Congenital dislocation of the knee, Proc Staff Meet., Mayo Clin 30 396, 1955

[22] Zadek, I · Congenital coxa vara, Arch Surg 30 62, 1935.

[23] LeMesurier, A B.. Developmental coxa vara, J. Bone & Joint Surg. 30B 595, 1948.

[24] Babb, F. S, Ghormley, R. K., and Chatterton, C C Congenital coxa vara, J. Bone & Joint Surg 31A·115, 1949.



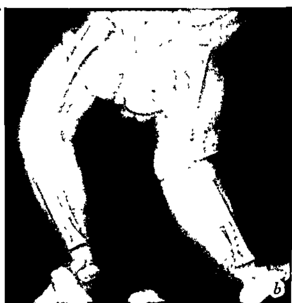


Fig 82 Congenital dislocation of the knee (Clayburgh, B. J.: Proc. Staff Meet. Mayo Clin. 30:396)

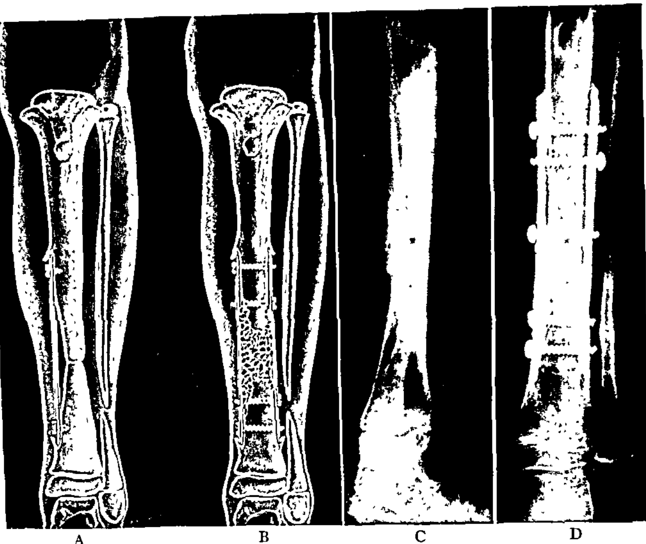


FIG. 85. Boyd dual graft for congenital pseudarthrosis. (A) First graft is held in position temporarily by 2 short screws. (B) Subsequently, both grafts are fixed in position by screws which pass through both grafts and intervening bone. Temporary screws have been removed and replaced by permanent screws. Trough filled with cancellous bone. (C and D) Views to demonstrate union and incorporation of the grafts. (Boyd, H. B.: J. Bone & Joint Surg. 23:497)

conditions identical with congenital pseudarthrosis result; (3) *congenital bowing of the tibia* with fracture resulting in pseudarthrosis; anterior bowing is more predisposed to this complication than posterior bowing.

Bony union is difficult and must be effected by a bone-grafting procedure. As a rule, the older the patient the more likely the success of union so that the age of puberty is selected by some surgeons as the appropriate time for surgery. However, by this time the limb is quite underdeveloped and short, and in many cases an artificial limb is better. When surgery is done in early childhood and is successful, sufficient time is left for growth factors to operate so that at puberty a well-developed, even normal, extremity results. Nothing is lost by failure of a well-planned operation in childhood. However, if surgery is postponed, further deformity of the extremity, particularly the anterior bowing and a concomitant calcaneovalgus deformity of the foot, should be prevented by wearing a leather lacer brace.

The principles of any bone-grafting operation to ensure success are: (1) secure internal fixation; (2) promote osteogenesis by adequate amounts of cortical and cancellous bone about the site of pseudarthrosis

The following is a typical procedure which has been used successfully by Boyd:[26] Two tibial cortical grafts and a supply of cancel-

[26] Boyd, H B · Congenital pseudarthrosis· treatment by dual bone grafts, J Bone & Joint Surg 23: 497, 1941.



FIG. 84. Congenital pseudarthrosis of tibia. Several unsuccessful attempts have been made to obtain union by bone grafting.

pression forces. A bone graft placed through the neck will aid in completely ossifying the neck. The operation is described briefly as follows:

Through a lateral incision, the femur is exposed subperiosteally below the great trochanter. A transverse osteotomy is performed, and the shaft is widely abducted so as to form a wide obtuse angle with the upper fragment. The position is held by a Smith-Petersen nail inserted in the head and the neck and attached at its outer end to a plate which has been affixed to the shaft. Instead of using a nail and a plate, a 1-piece metallic unit, such as a Moore or a Blount plate, consisting of a neck section and a shaft section, may be prepared preoperatively and bent to the appropriate angle and inserted at surgery. Postoperatively, a cast is unnecessary. Reossification almost inevitably occurs. The operation lengthens and thereby strengthens the hip abductors, and the Trendelenburg list becomes negative. Also, the femur is lengthened, and some of the limp due to shortening is overcome. However, the prognosis should be guarded inasmuch as recurrence can occur slowly regardless of any method, but most particularly during the growth period and when bone grafts alone are used.[25]

CONGENITAL PSEUDARTHROSIS OF THE TIBIA

In this condition, usually found at birth or shortly afterward, a section of the tibial shaft at the junction of the middle and the lower thirds is unossified and replaced by fibrous tissue which joins the upper and the lower remaining portions of the bone. The abnormal mobility suggests the term "pseudarthrosis," meaning a false joint. Examination of the infant reveals an underdeveloped lower extremity which is shorter than its fellow and usually bowed anteriorly. An indentation anteriorly over the site of the pseudarthrosis may be small as a dimple or may completely encircle the extremity as a constriction band. Roentgenograms demonstrate the lack of bone formation in the tibia at the junction of the middle and the lower thirds. The upper and the lower sections of the shaft are slender and densely sclerotic about the false joint. At a distance from the pseudarthrosis, these bones assume a normal size, shape and architecture. As a rule, the fibula is unaffected, although it may be bowed.

Three types are described. (1) *A defect in the bone* found at birth; (2) *a congenital bony cyst* through which fracture occurs, and the

[25] LeMesurier, A. B Correspondence to editor, J. Bone & Joint Surg. 33B:478, 1951.

the peronei and a posterior capsulotomy plus osteotomy of the tibia permit correction of the deformity. A prosthesis is worn until bone growth is completed. Then a lateral malleolus can be grafted to hold the talus in position. During the growth period, temporary epiphyseal arrest of the opposite extremity can be done to equalize lengths of the lower extremities. Failure to correct the foot deformity may necessitate a pantalar fusion, rarely amputation.

CONGENITAL ABSENCE OF THE TIBIA

Part or all of the tibia may be absent at birth. The affected extremity is shorter not only because the fibula is short but also because the fibula is displaced upward and lies alongside the lateral femoral condyle. The lower fibula lies lateral to the astragalus and the os calcis. The lack of tibial support allows the foot to swing medially into severe equinovarus. The entire leg deviates medially in relation to the longitudinal axis of the femur, and the fibula is bowed.

Treatment

The aim is to restore osseous continuity between the thigh and the foot. Surgery should be done as early as possible. The upper fibula is mobilized by cutting away the attachment of the biceps (after the common peroneal nerve is isolated and protected). Then it is placed in the intercondylar notch and held with sutures. It is immaterial whether a synostosis or a freely movable joint develops. At first the tight soft tissue structures will not allow the knee to be fully extended. However, this can be accomplished by successive casts. Finally, a leather lacer corset is fitted to the leg and holds the foot in extreme equinus, thereby overcoming the shortening and allowing the child to walk Constant walking exercise prevents atrophy and encourages bony and muscular development. Within a few years, when the lower end of the fibula and the astragalus and the os calcis are more ossified, the distal fibula is arthrodesed to the astragalus or, in its absence, to the os calcis, with the latter in extreme equinus. Weight-bearing in walking is on the toes. Over the years, the fibula will hypertrophy to several times its original size. The only alternative to this procedure is amputation.

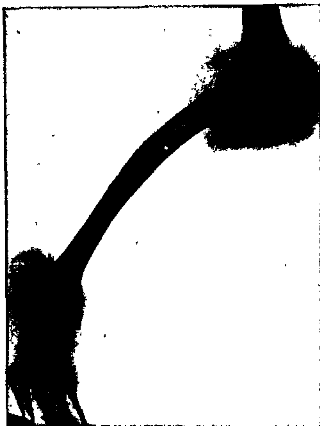


Fig. 87. Congenital absence of the tibia. The fibula is hypertrophied and dislocated at the knee. (Coventry, M. B., and Johnson, E. W., Jr.: J. Bone & Joint Surg. 34A:941)

CONGENITAL TORSION IN THE LOWER EXTREMITY

Torsion of a bone is defined as twisting or rotation on its longitudinal axis. The condition is found most commonly in the lower extremities, in the tibia or the femur. It is congenital or acquired. When present at birth, spontaneous correction occurs to a variable degree in the first few years in response to the stresses of walking and weight-bearing. The deformity which persists is compensated for by additional deformities at the hip, the knee, and the foot which permit forward progression of the foot in walking.

Tibial Torsion[28]

The angle which the median sagittal plane

[28] Kite, J. H : Tibial torsion, J. Bone & Joint Surg 36A.511, 1954.



Fig. 86. Congenital absence of the fibula. The lateral displacement of the foot at the ankle, anterior angulation of the tibia, equinus, and the frequently associated absence of digital rays are well shown. The patient displays the typical deformity of shortening of the leg and equinovalgus. (Coventry, M. B., and Johnson, E. W., Jr.: J. Bone & Joint Surg. 34A:941)

lous bone chips are removed from a donor or are supplied by the bone bank. The pseudarthrosis is exposed, and the fibrous tissue which may surprisingly extend into the surrounding soft tissue is completely excised. The eburnated bone is removed from the bone ends, and the medullary canal is opened up by drilling. To correct bowing, an Achilles tenotomy and a fibular osteotomy are required. If possible, the fibula should be preserved to aid the postoperative immobilization. The lateral surfaces of the tibia above and below are shaved down flat, and the tibial cortical grafts are affixed, one medially and the other laterally. A space is left between the ends of the tibial fragments into which are packed the cancellous bone chips. Only skin and subcutaneous tissue are closed. A cast is applied, and immobilization is continued until union is apparent. Fracture and nonunion are postoperative dangers, and protection of the leg by a leather lacer brace until puberty is mandatory.

CONGENITAL ABSENCE OF THE FIBULA[27]

The fibula is the long bone most frequently found absent at birth. Clinically, one finds anterior bowing of the tibia and dimpling of the skin at the distal third of the leg, marked talipes equinovalgus, absence of one or more rays of the foot, and absence of one or more, or fusion of two or more, tarsal bones. The concomitant failure of development of the calf and peroneal muscles throws abnormal stress on the tibia and the foot, resulting in the deformities. The extremity is shortened.

Treatment

Treatment consists of active weight-bearing and exercises with the aid of a prosthesis made of lightweight material such as celastic. At the age of 2 or 3, tenotomies of the Achilles and

[27] Coventry, M. B., and Johnson, E. W., Jr.: Congenital absence of the fibula, J. Bone & Joint Surg. 34A:941, 1952.

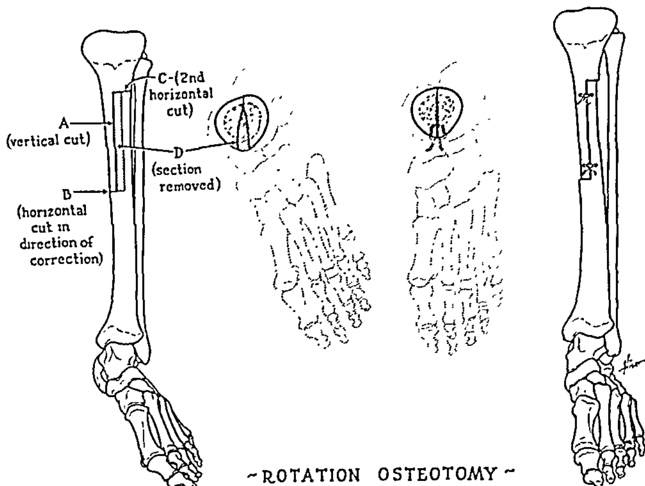


FIG. 89. Control rotation osteotomy of tibia (method of O'Donoghue). The distal horizontal limb of the Z-step-cut is always on the side of the direction of correction. (O'Donoghue, D. H.: South. M. J. 33:1145)

back in the "frog" or "spread-eagle" position. The limbs can be passively turned inward until the patellae face straight forward but not much farther. Rotation outward is excessive. Later, during the standing period, the feet turn outward and are usually flat. If the child turns the feet in order to point them forward, the patellae are rotated inward, affording a semilateral view of the knees which thus appear in valgus. The pseudo-knock-kneed condition is exposed by bringing the patellae to the forward position, which automatically eliminates the deformity. The mother's complaint, as a matter of fact, may be knock knees rather than flatfeet or an abnormal external rotation or "Charlie Chaplin" gait. Occasionally, actual knock knees may exist. These secondary deformities should always be sought and evaluated prior to treatment. Treatment consists of breaking faulty sleeping habits and continuously maintaining the feet in the inward position by bracing. Derotation is gradual over several months, but, as in acquired internal tortion, is limited by age of the patient. Persistent disabling deformity may require osteotomy.

Correction by Surgery. When torsion is associated with genu valgus or varus, a transverse osteotomy of the tibia is done just below the tibial condyles where union is most probable and displacement of the fragments less likely. The tibia and the fibula are sectioned, the distal tibia is rotated, and angulation is corrected at the same time as a cast is applied.

The *controlled rotation osteotomy*[29] is used when no varus or valgus correction is necessary. A Z-osteotomy is made through the upper third of the tibia. The distal horizontal limb of the Z-step-cut is made on the side of direction of correction. The proximal limb extends in the opposite direction. Between the

[29] O'Donoghue, D. H.: South. M. J. 33:1145, 1940.



FIG. 88. Internal tibial torsion. To test, the knees are flexed, and the ankle is held at a right angle. The relation of the foot to the sagittal plane measures the angle of torsion.

of the tibia forms with the coronal plane is known as the angle of torsion. When the sagittal plane is directly anteroposterior, both of the malleoli of the ankle should lie in the coronal plane, the talus faces forward, and the foot itself should point directly in front of the body. This angle is considered as 0° Normally, a small amount of external rotation is present and ranges from 0 to 40°, the foot pointing outward, and the malleolus displacing backward. When the distal tibia is twisted outward more than this amount in relation to the proximal tibia, it is considered an abnormal amount of external tibial torsion. When the distal tibia is twisted inwardly so that the foot points medial to the median plane, internal tibial torsion exists Measurement of the angle is determined by placing the patient in the sitting position, flexing the knee to a right angle, pointing the patella forward (which positions the proximal tibia), and placing the foot on the floor The angle is estimated by noting the deviation of the foot from the midposition.

Internal Torsion. When the *congenital type* is present, it is detected by the ease with which the extremity can be rotated passively externally and internally to the normal range as noted by the patella rolling out and in, respectively. The amount of angulation is chiefly at the distal end of the bone. When the child stands, in order that the feet be placed pointing forward, the patellae and the knees are rolled outward, giving a semilateral view of the knees. This simulates bowing of the knees. The pseudobowlegs are identified as such by facing the patellae forward, which automatically eliminates the deformity. Frequently, the mother's complaint may be bowlegs rather than pigeon toes. Occasionally, actual bowing may develop eventually. In such a case, the test of forward-pointed patellae fails to eliminate the deformity. Without treatment, the prognosis is poor. Spontaneous improvement rarely occurs. Treatment consists of holding the feet rotated continuously and externally by means of a Denis-Browne or a Fillauer splint, derotation taking place gradually over several months. The older the child, the less likely is the correction by conservative means. This is particularly true after the age of 6 or 7. If in such cases the deformity is extreme and disabling so that stumbling and falling is frequent, surgical correction by osteotomy is justified.

The *acquired type* of internal tibial torsion results from abnormal sleeping and sitting positions. The infant generally sleeps on his stomach in the knee-chest position with the toes turned in. On examination, the range of motion externally is limited, i.e , the patellae cannot be rolled out sufficiently. After sleeping habits are corrected, spontaneous derotation is usual. However, return to normal is expedited by bracing in the externally rotated position.

External Torsion. The anatomic outward twisting of the bone should not be confused with the outward *rotation* of the leg secondary to spastic paralysis or poliomyelitis. An overacting biceps femoris or a tight iliotibial band attached to the upper outer aspect of the leg will turn the leg outward, but actual torsion of the tibia is not present. External torsion of the bone results from faulty sleeping and sitting habits. The infant sleeps on its abdomen or its

the plantar concavity. This never should be interpreted as a flatfoot. Within the first year or two the pad disappears, and the true extent of the arch is revealed. In the normal foot, the talus, the navicular, the first cuneiform and the first metatarsal form a straight line as visualized from the lateral side. The *true criteria of a flatfoot* is a break in this line so that the *head of the talus points downward*, with or without the navicular. In the anteroposterior view, the *head of the talus points medially*, and the *navicular is displaced laterally* in relation to the talar head. The true extent of the displacement and the flattening of the longitudinal arch is revealed on weight-bearing. The *heel is everted* (valgus) so that the forefoot in relation to the hindfoot is really inverted. The examiner can restore the longitudinal arch to its normal appearance by passively inverting the heel and everting the forefoot. The condition of flatfoot is discussed further in the section on "The Foot."

Definition

Flatfoot is a congenitally developed condition in which the sole of the foot is convex, and the heel is in equinovalgus; the forepart of the foot is in extension; and the head of the talus forms the most prominent part of the sole. The European continentals call it "rocker foot," and the Americans usually reserve the term "flatfoot" for the milder degrees of the deformity.

Pathology

The talus points downward and medially, the calcaneus is in equinus, the navicular is displaced dorsally and laterally, and the cuboid is displaced dorsally. Both the Achilles tendon and the dorsiflexors are shortened. The dorsal capsules of the calcaneocuboid and the talonavicular joints are tight.

Etiology

The condition develops in the embryo stage along with other *congenital* deformities. It is often associated with spina bifida and arthrogryposis multiplex congenita (arthrogryposis). It may also be acquired from mistreatment of a clubfoot when a forceful attempt is made to overcome the equinus before the heel varus has been corrected.



Fig. 90. Congenital angulation of the tibia and the fibula.

Treatment

Conservative treatment by manipulation and corrective casts is inadequate. The following surgical methods have been used:

1. *Astragalectomy.* This is the most popular.
2. *Removal of head of the talus.*
3. *Soft-tissue operation.* The Achilles tendon is lengthened. The os calcis is pulled down by skeletal traction. A posterior tibiotalar capsulotomy is performed. Tight fascial bands are cut. All the dorsiflexor tendons are lengthened. Capsulotomies of the talonavicular and the calcaneocuboid joints are done. Then the dislocation is reduced at the midtarsal joints by plantar-flexing the forefoot, and a cast is applied with the knee held in flexion.
4. *Triple arthrodesis.* Disabling symptoms do not become manifest until adult life when the patient may first present himself for treat-

two transverse cuts, a wedge-shaped section of bone is removed from the anteromedial aspect of the tibia. The bone is osteotomized longitudinally on its posterior aspect, thereby freeing the 2 segments of tibia. The rotation is corrected by turning the distal fragment inward or outward, as the case may be, thereby approximating the edges anteriorly. Fixation of fragments is by sutures or transfixion screws.

Femoral Torsion

Internal torsion of the shaft of the femur occurs in congenital dislocation of the hip. In order that the patella may be directed forward, the entire bone is rotated outward, and the neck and the head of the femur are carried forward. This anteverted position of the neck is abnormal when it exceeds the normal 20° to 30° of anteversion. The forward placement of the head makes it difficult to retain the head in the acetabulum. In such cases, anteversion must be corrected by derotating the femur by an osteotomy. This is described in the section on "Congenital Dislocation of the Hip."

CONGENITAL DISLOCATION OF THE PATELLA[30]

The patella may be small and imperfectly developed at birth. It lies permanently in a laterally displaced position over the lateral or anterolateral aspect of the lateral femoral condyle. It is anchored in this position by the shortened fibers of the capsule and the vastus lateralis. The vastus medialis and the medial capsule are stretched The anterior portion of the lateral femoral condyle is flattened, but this may be a secondary change. Usually, the tibia is rotated externally, and genu valgus may or may not be present The abnormal lateral position of the patella causes pain and instability which in early childhood may be mild and cause no concern. However, with advancing age, the pain comes on with lesser forms of activity, and the weakness becomes so pronounced that the patient falls easily, and eventually crutches are required The imperfect mechanical situation causes flakes of cartilage to be knocked off into the joint where they develop into multiple loose osteocartilaginous bodies. Severe degenerative arthritis with narrowing of the joint space is the final result with passage of years.

The condition must be differentiated from the common recurrent dislocations of the patella occurring later and apparently without the hereditary factor.

Treatment

The procedures generally used in recurrent dislocation have not been entirely successful for the congenital condition. Conn's operation[31] gives satisfactory results. First, the external rotation and the valgus of the tibia are corrected by casts (only in the infant). In older individuals, the valgus and rotation need correction by a supracondylar osteotomy. Next, the knee capsule is exposed through a U-shaped incision which is deepened to include the synovium. The tendinous attachments of the vastus lateralis and the vastus medialis of the quadriceps mass are freed from the quadriceps tendon itself. The patella is pushed into its normal position in the midline, which leaves a diamond-shaped gap in the line of the lateral incision. A piece of similar shape (which conforms to a piece of tinfoil placed over this gap) is then cut from the redundant capsule on the inner aspect of the joint, which includes the capsule and the synovial membrane. This piece is transferred to and sutured to the margins of the opening in the lateral aspect. The medial opening is closed with sutures. The elongated fibers of the vastus medialis are shortened and sutured to the quadriceps and the upper border of the patella. The already short vastus lateralis is sutured to the quadriceps tendon. Weight-bearing is permitted in 3 weeks.

At a later age, removal of the patella and, if necessary, a knee "housecleaning" is done.

CONGENITAL FLATFOOT[32, 33]

(Congenital Rocker Foot)

The normal newborn foot does not possess a longitudinal arch, because a fat pad fills in

[30] Mumford, E B · Congenital dislocation of the patella, J. Bone & Joint Surg. 29 1083, 1947.

[31] Conn, H. R : A new method of operative reduction for congenital luxation of the patella, Boston Med & Surg J. 150:169, 1904

[32] Lamy, L , and Weissman, L · Congenital convex pes valgus, J. Bone & Joint Surg 21.79, 1939.

[33] Hark, F. W.: Rocker foot due to congenital subluxation of the talus, J Bone & Joint Surg. 32A·344, 1950.

D. The Spine

CONGENITAL TORTICOLLIS
(Congenital Wry Neck)

Torticollis is the deformity of tilting of the head toward one side and rotation toward the opposite side. It is generally caused by *muscle contraction* and/or shortened soft-tissue structures on one side of the neck, but other less common causes include *paralysis* of muscles on one side with resultant overaction of the muscles on the other side; *congenital deformities of cervical vertebrae; subluxation* of a cervical vertebra, usually spontaneous and occurring in children; *cervical adenitis* secondary to upper respiratory infection; *destructive cervical spine lesions; Sprengel's deformity; unilateral soft-tissue infection; neck tumors; myositis;* and a *central nervous system disorder*, particularly diseases of the basal ganglia. In any torticollis at any age, these causes must be considered in the differential diagnosis.

Etiology

The cause is unknown. The theories of causation include:

1. *Intra-uterine malposition.* Compression of vascular supply leads to ischemia and fibrosis of the sternocleidomastoid.
2. *Clotting in terminal veins* to the muscle, during labor.
3. *Tumor formation* in the sternocleidomastoid.

Pathology

At birth or within the first 2 weeks, a hard, *fusiform swelling within the sternocleidomastoid* muscle is found to consist of immature fibrous tissue, well demarcated from the surrounding normal-appearing muscle tissue. The "tumor" may occupy the entire thickness of the muscle, usually in the lower third. No evidence of hemorrhage or hemosiderin is found, so a remote possibility of trauma certainly cannot be substantiated. The fibrous mass subsides spontaneously, and the sternocleidomastoid becomes shortened and contracted, the process taking place over several months. Microscopically, the muscle throughout is deficient in muscle fibers, which are separated by mature fibrous tissue. The fibrous, unyielding shortening of the muscle

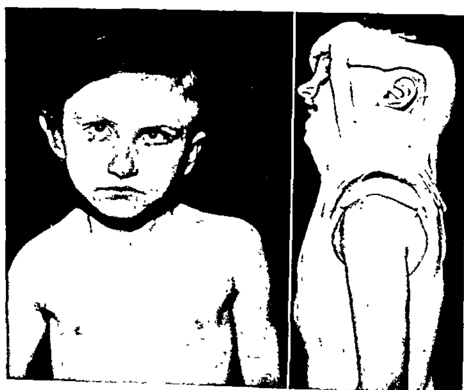


Fig. 91. Congenital torticollis (*Left*) Clinical appearance. Note the facial asymmetry and prominent sternocleidomastoids, particularly on the left side. (*Right*) Postoperative cast.

ment. The deformity is corrected by adequate resection of bone, and the calcaneocuboid, the talonavicular and the subastragalar joints are fused.

CONGENITAL ANGULATION OF THE TIBIA

(Congenital Bowing of Tibia; Congenital *Kyphoscoliotic Tibia*)

The tibia and the fibula may be bowed anteriorly or posteriorly and twisted internally or externally. This occurs at the junction of the middle and the lower thirds, at the site of origin of the primary ossification center and the usual area of predilection for pseudarthrosis. The degree of involvement varies from mild bowing of a well-developed tibial shaft to severe bowing and torsion in a tibia in which the bony architecture is deficient, thinned, and even cystic about the apex of the angulation. The cortex is thickest over the concave side of the curve. The remainder of the shaft and the epiphyses are normal. The skin is dimpled or retracted over the apex. A tendency to spontaneous fracture and subsequent pseudarthrosis exists. Occasionally, the growth in length of the limb is slightly retarded, although the epiphyseal plates seem to behave normally in appearance and time of fusion. When the bowing is anterior, talipes equinus is an associated deformity. The resistance of the clubfoot to treatment seems to parallel the severity of the bowing and its tendency to fracture and pseudarthrosis. Posterior and medial bowing is associated with severe talipes calcaneus. In this condition there is a tendency to shortness of the extremity and weakness of the calf muscles. However, there is no detectable abnormality of bony structure, no unusual tendency to fracture, and an excellent chance for spontaneous correction.[34, 35]

[34] Heyman, C. H., and Herndon, C. H. Congenital posterior angulation of the tibia, J. Bone & Joint Surg. 31A:571, 1949.

[35] Miller, B. F. Congenital posterior bowing of the tibia with talipes calcaneovalgus, J. Bone & Joint Surg. 31B:50, 1951.

Characteristics

The following are the main characteristics of this condition:[36]

1. Congenital anterior or posterior bowing, with rotation of tibia.
2. Location in distal portion of the middle third.
3. Site corresponds to site of origin of primary ossification center.
4. Thick ossification of cortex on concave side of curve; trabeculations are arranged radially from apex of curve.
5. Normal bone growth from epiphyseal ends.
6. Tendency to spontaneous correction.
7. Potential danger of pathologic fracture and nonunion. Pseudarthrosis more apt to develop in cases with anterior bowing.
8. Associated deformities:
   A. Hypoplasia—failure of development of a part; the fibula may be absent.
   B. Hyperplasia—partial gigantism as enlarged thumb.
   C. Syndactylism of hands or feet.
   D. Neurofibromatosis with café-au-lait spots.
   E. Dimpling of skin at apex of curved tibia.

Treatment

This should be cautious and conservative, even though the bony structure seems to be adequate and in spite of the angulation being posterior where the danger is supposedly slight. All cases should be regarded with respect. The infant should be constantly protected from injury until spontaneous correction of the curve and restoration of the bony architecture have occurred and the tendency to pseudarthrosis has lessened. This may require years. The temptation to osteotomize the tibia should be resisted. It is permissible to fit a long-length brace and apply a soft leather cuff over the apex which can be gradually tightened. Clubfoot deformities are treated with gentleness, avoiding a strain thrown above on the tibia during correction of equinus or calcaneus.

[36] Badgley, C. E., *et al.*: Congenital kyphoscoliotic tibia, J. Bone & Joint Surg. 34A:349, 1952.



Fig. 93. Klippel-Feil syndrome. Note short neck and low hairline. (Shoul, M. I., and Ritvo, M.: Clinical and roentgenological manifestations of Klippel-Feil syndrome, Am. J. Roentgenol. 68:369)

of the platysma and the deep fascia may also be severed. Some surgeons cut the tendinous origin of the sternocleidomastoid at the mastoid process, but this endangers the spinal accessory nerve. When this nerve traverses near the operative field, it should be identified and isolated and kept out of harm's way. Release of the soft-tissue structures should permit ample correction immediately. Postoperatively, manipulative stretchings are encouraged, and head traction is applied for several weeks. This maintains correction and strengthens the antagonist muscles. An alternative is to overcorrect the deformity at the conclusion of surgery and to apply a cast about the head and the chest (Calot Jacket). The cast is removed several weeks later, and manipulations and exercises are instituted.

KLIPPEL-FEIL SYNDROME

(Congenital Fusion of Cervical Vertebrae; Brevicollis)

Definition

This is a condition of congenitally fused and deformed cervical vertebrae, resulting in restricted neck motion and neurologic phenomena.

Pathology

Typically, several vertebrae are fused, usually in pairs, and the number of vertebrae is reduced, thereby shortening the spine in the

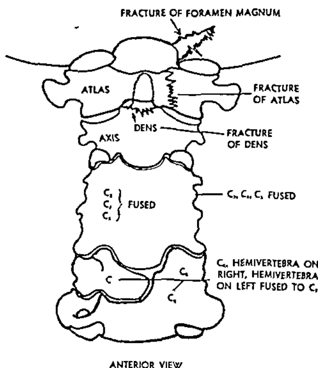


Fig. 94. Klippel-Feil syndrome. Diagrammatic representation of findings in cervical vertebrae at postmortem following accidental death. There are fractures of the foramen magnum, the atlas and the dens. The 3rd, the 4th and 5th cervical vertebrae are fused. The 6th cervical vertebra is in the form of 2 hemivertebrae, that on the left being fused to the 7th cervical vertebra (Shoul, M. I., and Ritvo, M.: Clinical and roentgenological manifestations of Klippel-Feil syndrome, Am. J. Roentgenol. 68:369)



FIG. 92. Klippel-Feil syndrome. Lateral view of cervical spine. The 1st cervical vertebra is elongated and tilted. The 2nd to the 6th cervical vertebrae are anomalous and fused. The normal cervical lordosis is completely absent. (Shoul, M. I, and Ritvo, M.: Clinical and roentgenological manifestations of Klippel-Feil syndrome, Am. J. Roentgenol. 68: 369)

permits neighboring structures to become contracted likewise, i e., the deep cervical fascia and the scaleni muscles. The face becomes asymmetrical and the head deformed secondary to the continued altered position over the years. The eyes eventually are situated at unequal levels. The cervical vertebrae over the growth years become deformed and adapted to the position. Thus it can readily be understood why the deformity becomes progressively fixed with passage of time.

CLINICAL FINDINGS

At birth or soon thereafter the infant's head gradually tilts to one side and rotates to the opposite side. The sternocleidomastoid muscle on the tilted side becomes taut and prominent and shortened. This is easily demonstrated on attempting passive correction of the rotation and the tilt. An elongated, indurated, nontender swelling appears in the lower third of the muscle in many cases. Occasionally, the circumscribed tumor is absent, but the muscle has a rigid, nonelastic feel to palpation. The deformity becomes progressively worse in degree and rigidity as the other soft-tissue structures and the cervical spine undergo adaptive changes. The head and the face eventually assume an asymmetrical shape, and a fixed scoliosis of the cervical spine results. In the adult, the deformity is practically uncorrectible.

TREATMENT

When treatment is instituted early, and the deformity is mild and readily overcome by passive movement, conservatism is adequate. The head is manipulated repeatedly toward the opposite position so that the sternocleidomastoid muscle is stretched. The daily stretchings are done by the mother with utmost gentleness. The child is placed in the crib in a strategic position of being forced to look and lift the head toward the direction of correction. For example, if the deformity is a right torticollis, the infant is positioned with its toys on the left side of the crib and the entrance to the room at the left so that he is forced to gaze toward the left at the person entering the room.

The severe and progressive cases should be determined early before rigidity and deformity of the face and the head set in. Surgery can be performed on infants as early as 3 months of age. Through a transverse incision just above the clavicle, the platysma muscle is split, and the sternocleidomastoid muscle is exposed. The sheath of the muscle is slit, and the tendinous insertions are cut at the medial end of the clavicle and the manubrium sterni. The muscle is allowed to retract. Chandler and Altenberg recommend removal of the fibrous tumor.[37] This can be shelled out and the muscle freed from underlying structures. It is questionable whether this step is necessary, inasmuch as the mass invariably recedes in a few months and does not seem to compromise the result. The clavicular attachment

[37] Chandler, F. A., and Altenberg, A.: "Congenital" muscular torticollis, J.A.M.A 125:476, 1944.



FIG. 95. Congenital absence of the sacrum and the coccyx. (Katz, J. F.: J. Bone & Joint Surg. 35A:398)



FIG. 96. Congenital absence of the sacrum and the coccyx. (Katz, J. F.: J. Bone & Joint Surg. 35A:398)

muscle, when necessary, and a double Z-plasty in the skin.

CONGENITAL ABSENCE OF SPINAL SEGMENTS[40, 41]

Agenesis of segments of the spinal column is rare and usually occurs in its terminal portions. Failure of development varies from the mere absence of the lower coccygeal segment to the complete nonexistence of vertebrae below the 10th thoracic segment. The extreme degrees of involvement are incompatible with life. Survival is possible only with absence of the sacrum or lesser degrees of the anomaly. The condition probably is due to some deleterious factor operative during the early weeks of fetal existence.

[40] Katz, J F: Congenital absence of the sacrum and coccyx, J. Bone & Joint Surg. 35A:398, 1953.

[41] Freedman, B.: Congenital absence of the sacrum and coccyx: report of a case and review of the literature, Brit. J. Surg 37:299, 1950.

CLINICAL PICTURE

When the sacrum is not present and the iliac bones meet posteriorly in the mid-line, the buttocks are flattened, the intergluteal fold is shortened, the sacrococcygeal prominence is lacking, and the transverse diameter of the pelvis is diminished. Neurologic involvement is prominent. There are atrophy and diminution of muscle power in the lower extremities and urinary and fecal incontinence. A characteristic cone-shaped appearance of the lower half of the body is apparent. The feet are deformed.

cervical region. The two vertebrae are joined by one continuous spinous process, and the bodies, if not joined by bony fusion, may be separated by a vestigial disk. The articulating processes are nonexistent, and the intervertebral foramina are narrowed and may encroach upon the nerve roots. Deformities of the cervical vertebrae, mainly of the bodies, result in scoliosis, kyphosis and torticollis which secondarily cause nerve root pressure and peripheral nerve symptoms. Spina bifida, particularly of the lowermost cervical vertebrae extending into the upper thoracic area, is frequent. Platybasia may be associated.

Clinical Picture

The main clinical characteristics are:

1. *Short neck.* The trapezei may be unduly prominent laterally and give a webbed appearance (webbed neck).
2. *Limited neck motion.* This particularly involves lateral bending and rotation.
3. *Lowered hairline.*
4. *Neurologic signs and symptoms.*[38] These are variable, depending on the degree of pathology. The nerve roots may be compressed by osseous malformation, scoliosis, kyphosis and torticollis, leading to signs of peripheral nerve irritation such as pain, burning sensations, cramps, etc.; or signs of actual nerve compression such as hypesthesia or anesthesia, weakness or paralysis, fibrillations, reduced deep reflexes, etc. When the spinal cord itself is involved, the lower extremities present signs of an upper motor lesion.

Roentgenographic Findings

Two or more vertebrae are found to be fused, particularly at their spinous processes, and are deformed. Oblique views demonstrate absence of articulations and the inadequacy of neural foramina. The pathology found varies from only 2 vertebrae, discovered on routine examination, which are asymptomatic, to involvement of all cervical vertebrae, severe malformations, curvatures, reduced number of vertebrae, and spina bifida.

[38] Mosberg, W H, Jr Klippel-Feil syndrome etiology and treatment of neurological signs, J Nerv & Ment. Dis. 117:479, 1953.

Importance of Condition

This lies mainly in distinguishing it from Pott's disease and congenital torticollis which require other forms of treatment. Deformity and rigidity of the cervical spine make it very susceptible to fracture from insignificant trauma such as the whiplash injury. Compression of nerve roots and the spinal cord itself is common following such trauma, even in the absence of definitely discernible fracture.

Treatment

Symptoms and findings of nerve root irritation, particularly when mild and of short duration, can frequently be relieved by traction and immobilization by a cast or a collar. However, neurologic phenomena which are resistant to conservative treatment and are disabling will require decompression of spinal cord and nerve roots. When one segment of the cervical spine is rigid, the segment immediately above suffers severe flexion strains, and disk rupture is probable. Its removal may be indicated. Loss of the disk will later eventuate in localized degenerative arthritic changes and spur formation which encroaches on the nerve root at the foramen. Decompression of this root is sufficient. Many cases are asymptomatic and require no treatment.

Correction of Deformity.[39] The shortening of the neck may be due to actual diminution in size or number of cervical vertebrae; or large cervical ribs and congenital elevation of the scapulae are often associated, accounting for an *apparent* shortening. The cervical ribs and first 3 thoracic ribs may be removed. This not only will produce apparent lengthening of the neck but also the upper thoracic vertebrae, when freed of their attachments, become more mobile, act more as cervical vertebrae and provide added flexor-extensor movement to the cervical spine. The scapulae are lowered surgically (see Sprengel's deformity). Occasionally, one encounters prominent folds of skin on each side of the neck (pterygium colli) associated with the Klippel-Feil syndrome, particularly in females. This is corrected by resection of fascia and part of

[39] Bonola, A Surgical treatment of the Klippel-Feil syndrome, J Bone & Joint Surg 38B 440, 1956.

fibrous band extends down to the deep fascia but not beyond so that the deep nerve and vascular structures are spared. However, the superficial lymphatics may be obstructed, resulting in marked swelling of the foot distally. This swelling is globular in appearance, completely enveloping the toes so that the latter appear to be absent. When the obstruction is overcome, the swelling subsides, and a more normal-looking foot is apparent.

Treatment is surgical. The band and the overlying skin are removed by a Z-plasty excision and suture of the skin, preferably in 2 stages, the anterior and posterior halves being done at separate times. If resection is extensive, skin grafting is necessary.

Ainhum

This is a condition affecting the 5th toe, less commonly the 4th, characterized by the *formation of a constriction band appearing* first on the plantar surface of the base of the toe and finally producing complete encirclement. Collagenic degeneration of the soft tissues and rarefying osteitis of bone develop distal to the point of constriction. It is most common in Negroes. Its congenital origin is controversial.

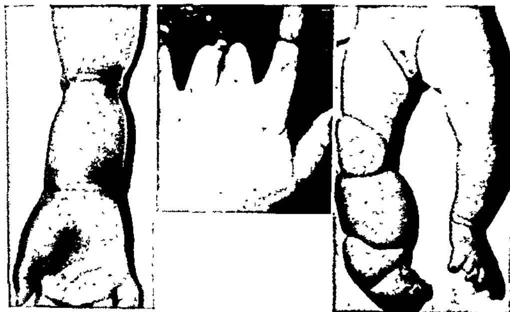


Fig. 98. Congenital constricting bands..These are attempts at segmentation. Swelling peripheral to the bands is due to lymphatic obstruction and will subside after removal of the constriction. (Blackfield, H. M., and Hause, D. P.: Congenital constricting bands of the extremities, Plast. & Reconstruct. Surg. 8:101)

E. General Defects

POLYDACTYLISM

Extra appendages may be present in the hand or the foot at birth. The appendage may vary in degree of development from a small skin tag to a completely formed finger or toe with phalanges and a metacarpal or a metatarsal. Usually, it consists of a small but recognizable digit containing phalanges and protruding from the lateral border of the hand or the foot and articulating with the distal end of the metatarsal or the metacarpal of the 5th toe or finger, respectively. It should be removed by disarticulation and cutting away the excess bone about the joint. When a fully developed digit is present, it is necessary to remove in addition the corresponding metacarpal or metatarsal. When doubt arises as to which of the digits is the accessory one, the digit with the least function is removed.

AMYOPLASIA CONGENITA

(Arthrogryposis Multiplex Congenita; Myodystrophia Fetalis)

This is a rare congenital condition displaying a marked hereditary tendency. The muscles of the extremities are aplastic, the muscle fibers being replaced by fibrofatty tissue. As a result, the joints upon which they act are very rigid. Clinically, these joints display limitation of both active and passive motion and usually are fixed in a position of flexion, adduction and inversion. In the foot the deformity is one of equinovarus which is extreme and unyielding, and a comparable deformity is found in the hand. The knees and the elbows present a fusiform appearance because of the muscle atrophy above and below. The muscles have a peculiar firm or rubbery feel, and the overlying skin is tense and glossy. The condition is painless. The spine and the back are practically never involved. The prognosis for overcoming the deformity is poor, but the life expectancy is not altered. Correction by conservative measures is valueless, because the factor of recurrence is strong. Improvement by operations on the bony structures, e.g., osteotomy or arthrodesis, should be done only if the patient's mental condition warrants it.



FIG. 97. Polydactylism. The proximal phalanx of the 6th or supernumerary toe articulates with the head of the 5th metatarsal.

The importance of this condition lies in its recognition. An innocent-looking clubfoot may prove to be impossible to cure because of an aplastic musculature. The typical signs of resistance to active and passive motion in all directions, symmetry of involvement, atrophic muscles, glossy skin and mental retardation should be sought in all cases of clubfoot and the unfortunate outlook made known. (See section on "Muscles.")

CONGENITAL CONSTRICTING BANDS

Well-defined transverse indentations of the skin and the underlying soft tissues completely encircling the extremity at one or several levels have the appearance of a band of tissue constricting the limb. The constriction varies in degree from a slight depression to a deep one simulating an embryonic attempt at amputation. Indeed, the disorder is frequently associated with absent parts of the extremities, e.g., fingers and toes. Other congenital anomalies as clubfoot are also found.

The lower third of the leg is the most common site for the band, where anterior or posterior bowing of the tibia may be present. The



Fig. 100. Osteogenesis imperfecta. Same case as in Figure 99. Trabeculations are better defined, and some transverse trabeculations are present. Cortices are thicker. Tendency to straightening of deformities with growth. This demonstrates progressive return to normal bony architecture and lessening tendency to fracture with advancing age.

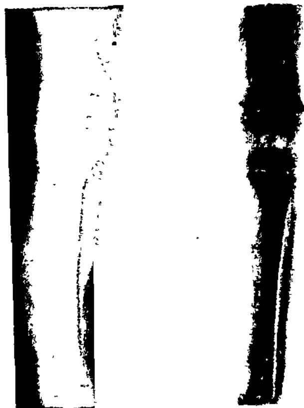


Fig. 101. Osteogenesis imperfecta, appearance later in childhood.

bone formation. Callus is abundant except in severe cases.

Associated Deformities. Vertebral bodies are translucent, shallow and biconcave as a result of disk indentation. Scoliosis is frequent. The skull is thin and globular. It may be crushed in severe cases at birth The pelvis may be compressed from side to side.

Clinical Picture

The condition may vary from mild to severe. The onset occurs at any time from before birth to late adolescence, rarely in adults.

Classification of Cases:

Fetal or Prenatal Form. Usually severe, many fractures throughout the body are present at birth. The skull feels like a membranous bag of bones. The baby may be a stillborn or die at birth or within a few weeks.

Infantile Form. This is less severe; multiple fractures occur; the skull is thin and globular and may resemble a hydrocephalic. If the child survives the first few years, the chance for lessening of the tendency to fracture and continued survival is good.

Adolescent Form or Osteogenesis Imperfecta Tarda. In this type the child is normal at birth, and fractures as a result of trivial trauma become manifest later in childhood. Later, the tendency to fracture is lost.

Characteristics. The following are the characteristics, one or all of which may be present in the patient and other members of his family:

Fractures. The number varies. In severe cases the fractures may be spontaneous. Pain is slight or absent. Healing readily occurs, usually with deformity which tends to lessen with continued longitudinal growth. The tendency to fracture lessens with advancing age.

Blue Sclerotics. This is a fairly deep indigo color, not merely the light blue seen in normal babies.

10

Developmental Conditions

OSTEOGENESIS IMPERFECTA[1-5]

(Fragilitas Ossium; Idiopathic Osteopsathyrosis; Periosteal Dysplasia)

Osteogenesis imperfecta is a hereditary condition characterized by fragility of bone, deafness, blue sclerotics, laxity of joints, and a tendency to improvement with age.

Etiology

The factor of *heredity* is demonstrable in many. Many prenatal cases are inherited as a mendelian recessive; the postnatal cases as a dominant. One or several of the characteristics of the condition may be present in other members of the family. The presence of blue sclerotics is said to be associated with hereditary types only.

Pathogenesis and Pathology

The primary defect is the *failure of formation of osteoblasts.* At the epiphyseal plate endochondral ossification proceeds normally as far as the stage of provisional calcification of cartilage. Very few osteoblasts appear, and osteoid formation is minimal. Some of the calcified cartilage may undergo direct metaplasia to bone. The formation of bone by periosteum is likewise deficient. The periosteum is thick, but the cambium layer is thin and relatively acellular. Occasionally, cartilage cells are formed at this site, perhaps representing persistence of the fetal histogenesis. In the skull *small scattered foci of bone formation and delayed closure of the fontanelles are characteristic.*

Grossly, the bones are shorter and thinner. The epiphyses in contrast with the shaft appear bulbous. Trabeculae are sparse, delicate and longitudinally disposed. No transverse trabeculae are seen. The cortex is very thin. Medullary contents are fatty or fibrous, rarely lymphoid. Deformity results from fractures and bending. The fracture often is subperiosteal and seems to heal mainly by periosteal



Fig. 99. Osteogenesis imperfecta. Marked osteoporosis, transverse trabeculations absent, longitudinal trabeculation poorly defined, thinned cortices, deformities at sites of healed fractures.

[1] Fairbank, Sir Thomas An Atlas of General Affections of the Skeleton, Baltimore, Williams & Wilkins, 1952.

[2] Luck, J. V.: Bone and Joint Diseases, Springfield, Ill, Thomas, 1950

[3] Wright, P. B, Gernstetter, S I, and Greenblatt, R B. Osteogenesis imperfecta therapeutic acceleration of bone age, J. Bone & Joint Surg 33A 939, 1951.

[4] Brailsford, J F Osteogenesis imperfecta Brit J Radiol. 16.129, 1943

[5] Weber, M: Osteogenesis imperfecta congenita, Arch Path. 9 984, 1930

Fig. 103. Chondro-osteodystrophy. Vertebrae are flattened and elongated anteroposteriorly. Kyphosis at dorsolumbar junction. Characteristic tongue-like process extends forward from the anterior aspect of the vertebral bodies. Ossification of apophyseal rings is delayed.



Fig. 104. Chondro-osteodystrophy. Delayed ossification of upper femoral epiphyses. Femoral head consists chiefly of a large amount of dystrophic cartilage which occupies the enlarged acetabulum. Femoral neck short and thick.

forward and sunk between the shoulders; and the gait is waddling. The facial appearance and intelligence are normal. A marked kyphosis is evident at the dorsolumbar junction. The chest is narrowed in the transverse diameter and elongated in the anteroposterior diameter (pectus carinatum). Although dwarfing is general, the spine is chiefly affected. All epiphyses are often enlarged and even bulbous. The joints, particularly the hips, are frequently stiff. Occasionally, ligamentous laxity with hypermobility occurs about the hands and the



FIG. 102. Chondro-osteodystrophy. Dwarfism, muscular weakness, crouched position, knock-knees, enlarged epiphyses, joint laxity, intelligent facies. (Case of J. F. Brailsford)

Deafness. Hearing is reduced in many by the third decade of life. Otosclerosis is frequently the cause.

Laxity of Joints. The susceptibility to strain and dislocation is common in families of the patients.

Feeble Musculature

Dwarfing caused by deformities of lower limbs and spine

Skull is broad, with prominent parietal and occipital bones (*crâne à rebord*)

Teeth Poorly Calcified. Permanent teeth are normal.

Blood Chemistry is Normal.

ROENTGENOGRAPHIC FINDINGS

The skeleton is osteoporotic, and the long bones appear elongate, thin, and possess thinned cortices and bulbous ends. The skull is thin, and wormian bones may be present. Vertebral bodies are translucent, shallow and biconcave, the disks being biconvex. Severe prenatal cases exhibit numerous fractures, particularly in the ribs, and the major long bones may be short, broad and thick.

TREATMENT

This consists essentially of protection of the child until the tendency to fracture has lessened. Adequate vitamin intake for bone deposition is necessary, but one must guard against excessive vitamin D medication by overzealous parents which would contribute to decalcification. The administration of estrogens and androgens may have a beneficial effect.

CHONDRO-OSTEODYSTROPHY

(Morquio-Brailsford Disease[6, 7, 8]

This rare condition is characterized by dwarfism, flattening of the vertebral bodies, marked kyphosis, defective ossification of many epiphyses, mainly in the spine and the hips, normal intelligence, and progressive weakness of the musculature. Familial tendencies are common. An early fatal outcome is usual.

CLINICAL PICTURE

The infant appears to be normal in all respects during the first 3 or 4 years. Gradually, a kyphotic deformity develops about the dorsolumbar junction; the neck appears shortened; failure to gain in height is noted; and the knees assume a valgus angulation. Because of muscle weakness, the child frequently supports himself by placing the hands upon the thighs. The typical appearance, as described by Morquio, is a round-backed, knock-kneed, flatfooted child who stands with the hips and the knees flexed in a crouching position; the neck is shortened, and the head is displaced

[6] Fairbank, Sir Thomas: An Atlas of General Affections of the Skeleton, Baltimore, Williams & Wilkins, 1952.

[7] Brailsford, J. F.: Chondro-osteodystrophy, J. Bone & Joint Surg. 34B:53, 1951

[8] Morquio, L. Sur une forme dystrophie osseuse familiale, Arch de med d. enf 32.129, 1929; 38·5, 1931.

by the spine; the multiple ossific centers in the epiphysis are mulberrylike; several small centers surround one main center; the epiphysis becomes flattened; muscle power is normal; hereditary and familial influences are common; the tendency is toward improvement; and life expectancy is normal. Dwarfing is not as severe.

DYSPLASIA EPIPHYSIALIS MULTIPLEX[9]

This is a rare, often hereditary, developmental defect characterized by abnormal ossification of many epiphyses and stunting of growth, which, contrary to Morquio's disease, is not chiefly caused by restriction of growth of the spine. The tendency is toward improvement, the epiphyses being moderately malformed, and degenerative arthritic changes in the adult are usual. The musculature is unaffected.

Clinical Picture

Pain and stiffness in the knees and the hips, difficulty in walking, and restriction of motion at the shoulders are early complaints. The hands present short, thick fingers. Most commonly the hips, the shoulders, the knees and the ankles are involved. The spine may be affected, but kyphosis is unusual. No deformity is apparent other than short stature. In the adult, bilateral symmetrical osteoarthritic changes common in the hips should suggest x-ray studies of other joints. Several members of a family may be affected. Life expectancy is normal

Roentgenographic Findings

The centers of ossification are late in appearing, are multiple and often arranged in a mulberrylike fashion about one main central nucleus. Eventually, the epiphysis becomes normal in density, and fusion of the epiphyseal line occurs at the usual time. The epiphysis is smooth in outline but flattened, often with a bony prolongation inferiorly. The joint space becomes narrowed, and degenerative changes supervene in the adult. The glenoid and the acetabulum, at first normal, eventually conform to the shape of the epiphysis.

[9] Fairbanks, Sir T.: Dysplasia epiphysialis multiplex, Brit J Surg. 34.225, 1947

A characteristic deformity at the ankle consists of the lower tibial epiphysis diminishing in depth from within outward so that the joint surface is oblique. The shafts of the long bones are shorter than normal. Carpal and tarsal bones ossify late.

The spine, when involved, shows only delayed ossification and fragmentation of the epiphyseal plates with wedging of the bodies, but the deformity is never as severe as in Morquio's disease.

DYSPLASIA EPIPHYSIALIS PUNCTATA[10-13]

(Chondrodystrophia Calcificans Congenita; Stippled Epiphyses)

This is a rare congenital condition characterized mainly by discrete spots of calcification affecting cartilaginous structures. It is not hereditary and rarely familial.

Pathology

Throughout the entire body multiple focal areas of calcification develop in cartilaginous precursors of bones. Interspersed between calcified foci are patchy areas of mucoid and cystic degeneration. The spots of calcified cartilage are found especially in epiphyses where they tend to concentrate adjacent to the metaphyses. The long bones are shortened, and the diaphyses are widened at their metaphyseal ends. If the patient survives beyond infancy, the epiphyses become completely calcified and subsequently ossified.

Clinical Picture

The subject is usually stillborn or seldom survives infancy. The rare case living beyond childhood develops shortness of the affected

[10] Conradi, E · Vorzeitiges Auftreten von Knochen —und eigenartigen Verkalkungskernen bei Chondrodystrophia fotalis hypoplastica, Jahrbuch fur Kinderheilkunde 80·86, 1914

[11] Fairbank, H. A. T · General diseases of skeleton, Brit. J. Surg. 15 120, 1927.

[12] Frank, W. W., and Denny, M. B . Dysplasia epiphysialis punctata, J. Bone & Joint Surg 36B:118, 1954.

[13] Karlen, A. G , and Cameron, J. A. P.: Dysplasia epiphysialis punctata, J. Bone & Joint Surg 39B·293, 1957



FIG. 105. Dysplasia epiphysialis multiplex, adult appearance. Epiphysis is dense and flattened, the glenoid conforming to its shape Narrowing and degenerative changes which characteristically and early restrict shoulder motion. Findings are bilateral, symmetrical, and occurring in large joints, especially hips, shoulders, knees and ankles.

feet. The fingers are usually broad and blunt Pain is unusual.

The course is for the dwarfing, the deformities and the muscle weakness to become progressively more marked. Survival beyond childhood is extremely rare

ROENTGENOGRAPHIC FINDINGS

Characteristic findings occur in the spine and the hips The vertebrae are flattened and are elongated anteroposteriorly The superior and the inferior margins are irregular and ill-defined. Anterior wedging is most marked at the cervicodorsal junction, which causes the shortened neck, and at the dorsolumbar junction, which creates a kyphosis. A characteristic tonguelike process extends forward from the anterior aspect of the vertebral bodies Ossification of the epiphyseal rings is delayed and irregular. The disks are unusually wide.

The upper femoral epiphysis displays delayed ossification with formation of multiple irregular centers Eventually, the femoral head becomes irregular, flattened and fragmented. The femoral neck appears short and thick. A coxa vara deformity is common. The acetabulum is quite large and irregular. The joint space appears unusually wide since it is occupied by a large amount of dystrophic cartilage. Occasionally, dislocation is a consequence of extreme muscle flaccidity.

Similar changes are seen in other epiphyses The result is a large epiphysis, irregularly ossified, and a shortened diaphysis.

DIFFERENTIAL DIAGNOSIS

Rickets is simulated by the swollen joints. However, rickets displays generalized osteoporosis, mainly in the diaphyses. The epiphysis is ossified from a single central nucleus Shafts of weight-bearing bones become bent. Genu varum is far more common. In Morquio's disease, knock knees are typical. Fractures are frequent. The condition responds to Vitamin D.

Achondroplasia. The spine is normal in length, and the limbs in contrast are very short Muscle power is unusually well developed. Gross epiphyseal and articular changes are never seen, because epiphyseal growth takes place from a single nucleus which progresses to maturity.

Gargoylism. This chondro-osteodystrophy type of dwarf is characterized by mental deficiency, heavy facies, cloudy corneae and enlargement of the liver and the spleen. The shapes of the vertebral bodies are different.

Hypothyroidism. A generalized delay in ossification is typical Epiphyseal centers appear late, are multiple, and become deformed with weight-bearing. The condition responds to thyroid medication, the multiple nuclei fuse into one well-formed nucleus

Dysplasia Epiphysialis Multiplex. This simulates Morquio's disease However, stunting of growth is general and not chiefly caused

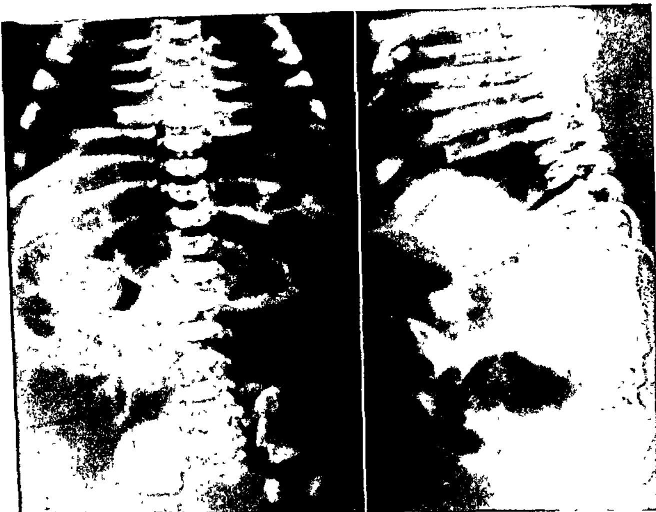


Fig. 107. Osteopetrosis. (N. U. Case #294)

white, is solid on cross section and possesses club-shaped extremities due to widened metaphyses. If the process is intermittent, transverse bands of dense bone alternate with bands of normal bone throughout the shaft. In the epiphysis, concentric alternating rings of dense and normal bone are often formed. The vertebral body typically forms a transverse band of dense bone at either extremity with an intervening area of normal bone. Skull diploe are fused as one. The air sinuses are often replaced by dense bone or are entirely absent. The pituitary fossa is shallow, and the posterior clinoid processes are clubbed and encroach upon the fossa. Bony enlargement narrows the skull foramina and compresses nerve structures, particularly the optic nerve Peculiarly, the mandible is immune.

Clinical Picture

The condition starts during gestation and is progressive until longitudinal growth stops. The intensity of the disorder varies. In a mild type, formation of dense bone occurs slowly, intermittently and incompletely, and the patient survives. The more severe malignant type often occurs when consanguinity of parents exists. All bones early and rapidly become very dense, devoid of architecture, and usually intensely hard or chalky and brittle. Fractures are frequent and heal slowly. The fracture is transverse and sharply abrupt. An anemia develops because of bony or fibrotic replacement of the marrow. Its severity depends upon the capacity for extramedullary hemopoietic tissues to undergo compensatory hypertrophy. Optic atrophy, facial or ocular palsy, deafness and hydrocephalus are complications.

Laboratory Findings

Serum calcium, phosphorus and phosphatase are normal.

extremities. The main characteristics of the disease are:

1. *Flexion deformities* of joints, especially knees and elbows. The cause is probably muscular and capsular fibrosis.

2. *Bilateral congenital cataracts* are found in most cases.

3. *Dwarfism of short-limb type* as in achondroplasia Femurs and humeri are chiefly involved. A single limb may be affected.

Roentgenologic Findings

Opaque, discrete or coalescing dots occupy cartilaginous structures such as the epiphyses, carpals and tarsals, cartilaginous portions of ribs, iliac apophyses, etc. These appear earlier than the usual time of appearance of ossification centers One or more long bones may be shortened, thickened and bowed. The metaphyses of the shortened bones are splayed, and their epiphyseal borders are irregular.

Differential Diagnosis

The condition must be differentiated from dysplasia epiphysialis multiplex and cretinism.

OSTEOPETROSIS[14, 15]

(Albers-Schonberg's Disease, Marble Bones; Chalk Bones)

This is a rare developmental abnormality in which the bony structure throughout the body becomes increasingly dense and brittle. Complications are caused by obliteration of the marrow (anemia, hemorrhages) and by bony encroachment on skull foramina (optic atrophy, deafness).

Pathology

The process appears to be continued deposition of new bone on unresorbed calcified cartilage or primary spongiosa A failure of remodeling results in marked widening of the metaphyses and a club-shaped appearance of long bones. The condition starts before or at birth and continues uninterruptedly or intermittently until growth stops. The bone grossly is grayish white on section. It may be as hard as marble or have the consistency or brittleness of chalk. The medullary cavity is obliterated. Microscopically, the trabeculae are greatly increased in number and thickness and appear to be disorganized. Haversian canals are rare. Many islets of hypercalcified cartilage persist among the dense bony trabeculae. Marrow spaces are small, infrequent and fibrotic. Osteoblasts are normal or increased in number. Osteoclasts are normal or absent.

The typical long bone is very dense and

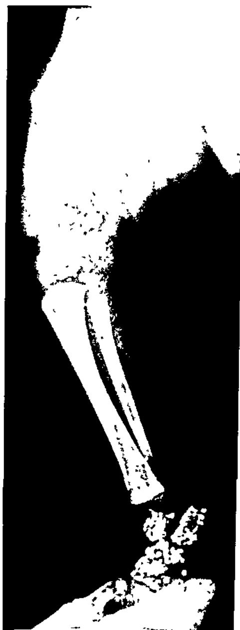


Fig. 106. Dysplasia epiphysialis punctata. (*Karlen, A. G., and Cameron, J. A. F.: J. Bone & Joint Surg. 39B: 293*)

[14] Fairbank, T.· An Atlas of General Affections of the Skeleton, Baltimore, Williams & Wilkins, 1952.

[15] Albers-Schonberg, H: Röntgenbilder einer seltenen Knockenerkrankung, Munchen Med Wchnschr. 51:365, 1904; Fortschr. Geb. Röntgenstrahlen 11 261, 1907.



FIG. 109 (*Left*). Melorheostosis of the tibia. The structureless dense streak which extends beyond the surface in undulating fashion resembles "candle drippings."

FIG. 110 (*Right*). Melorheostosis of the tarsals and metatarsals. The dense blotches are most apparent in the scaphoid and first cuneiform.

show a central focus of dense or normal bone surrounded by alternating rings of increased or normal density.

MELORHEOSTOSIS[16]

(Leri Type of Osteopetrosis; Monomelic Flowing Hyperostosis)

Melorheostosis is a rare condition in which dense bone formation in a bone resembles the flow of candle drippings.

ETIOLOGY

The cause of this disease is unknown. Heredity plays no part. Suggested theories include: (1) ischemia secondary to sympathetic system disturbance, and (2) a developmental error.

CLINICAL PICTURE

Age. Begins in childhood.

Area of Predilection. The lower more often than the upper extremity. Typically, the changes are confined to one limb.

Symptoms and Signs. The following are characteristic:

Pain, dull and aching, seldom severe, likely in older patients.

Limitation of motion in joints of affected limb.

Deformity of bone due to irregular thickening.

Shortening of the limb in some cases.

Swelling, edema, induration, coldness, increased perspiration and *skin changes* (scleroderma) suggest sympathetic ischemic phenomena. The resulting fibrosis of muscles and other soft tissues is responsible for stiffness and limitation of motion.

ROENTGENOLOGIC FINDINGS

A dense structureless streak or blotch of part of a bone resembles the flow of radiopaque substance. The surface is undulated. The scapula or half pelvis corresponding to the affected limb usually shows dense patches. The skull, the spine and the ribs invariably escape.

PATHOLOGY

Microscopically, there is a compact overcrowding of lamellae arranged in a bizarre

[16] Fairbank, Sir T.: An Atlas of General Affections of the Skeleton, Baltimore, Williams & Wilkins, 1952.



PLATE 11. Marble bone (× 85).

ROENTGENOLOGIC FINDINGS

The entire long bone, including the epiphyses, may be uniformly dense and completely devoid of structure. Occasionally, less dense areas of normal bone are interspersed. When the process is active temporarily, a dense band forms in the metaphysis and with continued growth displaces distally along the diaphysis. The metaphyses are chiefly affected, becoming clubbed; the enlargement ends abruptly at its junction with the diaphysis. The ilia often show alternating dense and clear curved bands parallel with the crests. Density of the skull is greatest at the base. The pituitary fossa is small, and the thick clubbed posterior processes close in on the fossa. Air sinuses are absent or dense. The maxilla is affected, but the mandible almost invariably escapes. Each vertebra displays dense bands at either end and a clear band between. An epiphysis may



FIG. 108. Melorheostosis. The peculiar streaked sclerosis of bone resembling candle drippings is well demonstrated. Frequently associated with congenital neurofibromatosis. (McCarroll, H. R.: J. Bone & Joint Surg. 32A:601)

Roentgenographic Findings

Cartilage-filled clear spaces of varying size and shape appear in the metaphysis and tend toward a columnar arrangement. The columns are separated by septa of increased bony density which cause a striated or streaky appearance. The metaphysis is irregularly expanded. The diaphysis is short and thick, occasionally curved. The large mass of cartilage may bulge and interrupt the continuity of the cortex. The epiphysis adjacent to an involved metaphysis often displays dense mottling. In the phalanges all semblance of the original bone may be lost. Ilium involvement is characterized by cartilage columns which radiate in a fanlike manner toward the crest. As adolescence approaches, the cartilage columns and spaces become less well-defined with dense spots appearing throughout the metaphysis, suggesting replacement by ossification.

Treatment

Occasionally, osteotomy is necessary to correct deformity. The fragments readily unite. Epiphyseal arrest in the opposite extremity combats limb inequality.

Mafucci's Syndrome

This consists of dyschondroplasia associated with cavernous hemangiomata and phleboliths in the soft tissues.



Fig. 112. Dyschondroplasia. Cartilage-filled clear spaces of varying size and shape, columnar arrangement, density of bone between cartilage columns is increased, metaphyseal enlargement, thickened shaft, shortening of bone, mottled epiphyses.

HEREDITARY MULTIPLE EXOSTOSES

(Hereditary Deforming Dyschondroplasia; Metaphyseal Aclasis)

In this condition many osteocartilaginous exostoses form at the metaphyses of long bones. Several members of a family are usually affected.

Etiology

The cause is unknown. The following theories of pathogenesis have been advanced:

1. **Failure of Development of the Periosteum.**[19] The cortex fails to attain full thickness, and the inadequate periosteum cannot restrict the outward growth of bone.

2. **Failure of Osteoclastic Activity.**[20] The modeling and shaping process of bone is deficient, explaining the widened trumpet-shaped appearance of the metaphysis. If this theory were correct, *metaphyseal aclasis* should be applied to this condition.

3. **Deficient Perichondrium, Permitting Overgrowth of Cartilage.**[21] The cartilage becomes transformed into bone. Because all displaced cartilage is prone to malignant change, this explains the occasional malignant degeneration of an exostosis.

[19] Geschickter, C F, and Copeland, M M.: Tumors of Bone, ed 3, p 79, Philadelphia, Lippincott, 1949.

[20] Keith, Sir A : J Anat. 54:101, 1920.

[21] Bennett, G. E, and Berkheimer, G. A.· Surgery 10.781, 1941.

manner and an interlacing of immature and adult bone.

Treatment

Sympathectomy may effect relief of pain, appearance of warmth and dryness, and increase of motion. The effect should be tested presurgically by sympathetic block.[17]

DYSCHONDROPLASIA[18]

(Ollier's Disease; Multiple Enchondromata)

Dyschondroplasia is a rare developmental condition characterized by large rounded masses or columns of cartilage in the metaphyses of certain bones, particularly the long bones. The cartilage proliferates normally at the epiphyseal plate but fails to become calcified, resorbed, or replaced by newly formed bone. This results in accumulation of large amounts of cartilage in the metaphysis and reduction of longitudinal growth. The cartilaginous masses bulge the metaphysis and may penetrate the cortex with formation of ecchondromata. Dense septa of normal bone divide the cartilage into lobules. The diaphysis is short and broadened. Chiefly involved are the long bones formed in cartilage, particularly at the more rapidly growing ends, e.g., about the knee and at the distal ends of the radius and the ulna. The condition is predominantly unilateral In the small long bones of the hands, the enchondromata occupy the entire bone and grow to large size, seriously deforming the hand. Microscopically, the masses consist of hyaline cartilage with cells of varying size irregularly arranged. Some calcification and replacement by ossification is seen in older patients approaching adolescence

Clinical Picture

Age. The patient is normal at birth; the condition becomes manifest at any time during the growth period.

Area of Predilection. Long bones especially at actively growing ends. Knee, distal ulna and radius, upper end of humerus. Long bones of hands and feet, especially the phalanges. Pelvis, particularly the iliac crest.

Symptoms and Signs. The affected extremity is shortened. Deformities: valgus or varus result when growth is retarded unequally at the epiphyseal line; the ulna is shortened more than the radius, which in consequence becomes bowed and dislocated at the radial head; large enchondromata of the phalanges cause gross deformity of the hand. Fractures are uncommon except in the hand.



Fig. 111. Melorheostosis. Dense, structureless, cortical and endosteal, flowing hyperostosis resembling candle drippings.

[17] Hess, W E, and Street, D M Melorheostosis, relief of pain by sympathectomy, J. Bone & Joint Surg 32A 422, 1950

[18] Fairbank, Sir T.: An Atlas of General Affections of the Skeleton, Baltimore, Williams & Wilkins, 1952

Prognosis

Ordinarily, these are benign growths, but the possibility of malignant change must be entertained. Growth of the exostoses is discontinued when skeletal growth stops.

Treatment

The exostosis is removed when it is constantly subjected to injury, causes deformity, compresses important structures or is suspected of undergoing malignant transformation.

ACHONDROPLASIA

(Chondrodystrophia Fetalis; Micromelia)

Achondroplasia is a congenital developmental condition characterized pathologically by defective endochondral ossification, affecting chiefly the long bones, and clinically by a peculiar dwarfism in which the extremities are shortened, whereas the trunk remains relatively unaffected.

Etiology

The actual cause is unknown. Hereditary tendencies are occasionally apparent, the condition being manifest in half the children of parents, one of whom is an achondroplastic dwarf. Females are more often affected.

Pathology

The process of endochondral ossification at the epiphyseal growth plates is disturbed. Instead of orderly proliferation, palisading and formation of scaffolds of calcified cartilage, the cartilage is degenerate and very vascular. Therefore, ossification for longitudinal growth is greatly retarded. On the other hand, periosteal ossification proceeds normally, and bone diameter is assured. This form of ossification may even be excessive, particularly at the metaphysis where the cortex flares outward and appears to embrace the epiphysis. Within the epiphysis itself, where palisading and provisional calcification are unnecessary, ossification proceeds at a normal pace.

Although all bones dependent upon endochondral ossification are involved in the process, the long bones of the limbs are chiefly affected. The base of the skull, which develops in cartilage, is compromised by premature fusion, forming a single mass of bone. Growth is retarded, the base remaining short while the rest of the skull grows normally.

Clinical Picture

Dwarfism is often apparent at or soon after birth. Prematurity is frequent, most of the newborn being stillbirths or failing to live beyond the first year of birth. Those that survive become healthy and robust, and life expectancy is normal.

During childhood, the extremities lag behind in longitudinal growth so that, although the spine is affected to some extent, the limbs appear strikingly short in comparison with the trunk. The hands present a typical "trident" appearance. They are short and broad, the fingers being short, thick and diverging. Be-



Fig. 114. Achondroplastic dwarf.

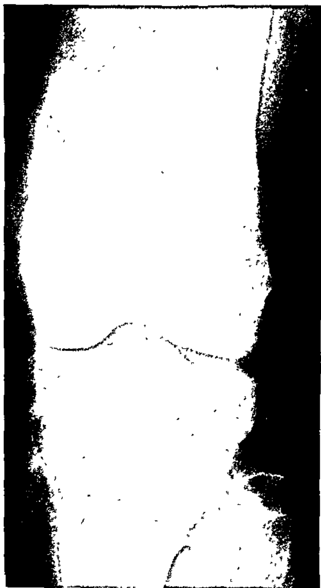


Fig. 113. Hereditary multiple exostoses. The metaphyses are trumpet-shaped and poorly trabeculated. The exostoses which point away from the ends of the bones are actually much larger than is represented on the roentgenogram because they are capped with thick hyaline cartilage.

Pathology

Only bone arising in cartilage and eventually enclosed by a sheath as subperiosteal bone is affected. Membranous bone is immune. Therefore, exostoses develop at the growing ends of long bones and especially where such growth is most active, i.e., the distal end of the femur and the radius and upper end of the tibia and the humerus. The metaphysis is uniformly enlarged, has parallel sides and, on approaching the central part of the shaft, abruptly narrows to the normal diameter, a configuration which Keith calls "trumpeting." Irregular projections extend from the surface and point away from the end of the bone. They may be conical, spiked, or globular in shape. These exostoses first occur near the epiphyseal cartilage and are displaced along the shaft with growth. One can almost estimate their time of appearance by measuring the distance from the epiphyseal line. Growth in length of the bone is interfered with as the cartilage grows outward so that, for example, if the fibula lags behind the tibia, the latter may become curved.

Microscopically, the metaphysis shows poor trabeculations, which are continuous with those within the central portion of the exostosis. Jaffe describes the exostosis as an outpouching of the cortex.[22] The surface is a zone of hyaline cartilage with active endochondral ossification beneath it. When growth stops, the surface has a thin layer of nonproliferating cartilage resting on a thin layer of bone. Underneath is spongy bone with delicate trabeculae, fatty marrow and islands of calcified cartilage.

Clinical Picture

The patient may or may not be of shortened stature, depending upon the severity of the process. Irregular hard prominences near the ends of long bones may be visible or at least palpable. These may be tender; the overlying skin, if subjected to pressure or friction, is tender, reddened and swollen. Numbness, paresthesias and muscle weakness may result from nerve pressure. Involvement of adjacent tendons restricts movement. The excrescences may be fractured but unite readily. When an asymptomatic exostosis suddenly without cause becomes enlarged and painful, malignant degeneration should be suspected.

Roentgenographic Findings

These reveal the trumpetlike metaphysis with very little compact cortical bone, continuous with that of the exostosis. The paucity of normal trabeculae causes a relatively less dense appearance of the interior of the growth.

Complications

Complications include fracture, overlying bursitis, pressure on tendons and nerves, shortening and bowing of the extremities, and sarcomatous degeneration.

[22] Jaffe, H. L.: Arch Path. 36:335.

ENGELMANN'S DISEASE[23-26]

(Progressive Diaphyseal Dysplasia; Progressive Diaphyseal Hyperostosis)

This syndrome is characterized by symmetrical fusiform enlargement and sclerosis of the shafts of major long bones associated with similar hypertrophic dense changes in the skull. The etiology is unknown.

[23] Camurati, M.: Di un Raro Caso di Osteite Simmetrica Ereditaria degli arti Inferiori, La Chir. degli Organi de Movimento 6:662, 1922.

[24] Engelmann, G.: Ein Fall von Osteopathia hyperostotica (sclerotisans) multiplex infantilis, Fortsch. Geb. Rontgenstrahlen 39.1101, 1929

[25] Fairbank, T.: An Atlas of General Affections of the Skeleton, Baltimore, Williams & Wilkins, 1952.

[26] Griffiths, D. LL.: Engelmann's disease, J. Bone & Joint Surg. 38B:312, 1956.



Fig 116. Engelmann's disease. The cortical thickening involves the diaphyses, ends abruptly at the metaphyses and encroaches upon the medullary canal. (Gillespie, J. B., and Mussey, R. D.: J. Pediat. 38:55)

Clinical Picture

The condition is first observed in early childhood. Sexes are equally affected. Infants walk late, and dentition is retarded. Often the patient is gaunt and thin, possessing poor muscle tone, and fatigues easily. The diaphyses of long bones are *often palpably thickened*. Soon or late, usually before the age of 7, aching pain in the legs becomes a prominent com-



Fig. 117. Engelmann's disease. (Griffiths, D. LL.: J. Bone & Joint Surg. 38B:312)



FIG 115. Osteopoikilosis.

cause the humeri and the femora are intensively involved, shortening affects the arms and the thighs more than the forearms and the legs *The fibula is relatively unaffected in contrast with the tibia.* As a result, the head of the fibula appears prominent and lies at a high level on the lateral aspect of the knee joint.

The musculature is well developed, and often muscle power is superior, enabling these individuals to perform feats of strength and acrobatics.

The hip joints lie posterior to the central axis of the pelvis. Consequently, the pelvis tilts forward, the buttocks are prominent, the sacrum lies in a horizontal plane, lumbar lordosis is exaggerated, and a compensatory increase of the thoracic kyphosis develops. The peculiar rolling gait has been attributed to hip displacement and pelvic tilting.

The head is brachycephalic. The forehead is prominent, and the bridge of the nose is depressed.

The chest is small and flat, the ribs being abnormally short.

Intelligence and sexual development are normal.

Roentgenographic Findings

The long bones are short and, because of reduction in length, are apparently rather than actually increased in diameter. The ends of the shafts are splayed. The epiphyseal border of the metaphysis is often indented at its center. The epiphyseal ossification center, which is not dependent upon endochondral ossification, is well developed and well circumscribed. It lies in close apposition to the metaphysis, fitting snugly into the V-shaped indentation. This displacement of the center away from the joint gives the illusion of marked widening of the joint space. The clavicles and the fibulae are less affected by shortening than the other long bones. The ribs are short and the sternum is broad and thick. The scapula is rectangular and small.

The pelvis is reduced in all dimensions. The ilium presents a characteristic rectangular appearance. The roof of the acetabulum is flat and *horizontally disposed. The hip joint lies* farther back than usual, and the acetabulum is close to the sacrosciatic notch. The sacrum is narrow and horizontally disposed.

The skull is large. Premature fusion of the basal centers considerably shortens the base of the skull, the foramen magnum remaining small and funnel-shaped. The facial bones are not involved.

In the spine, endochondral ossification seems to be relatively unaffected so that, although the depth of the vertebral bodies is reduced, the total length of the spine is much less diminished than that of the extremities.

OSTEOPOIKILOSIS

(Osteopathia Condensans Disseminata)

This condition is characterized by the development of multiple dense spots in many bones. These appear during the growth period, are asymptomatic and persist throughout adult life. The affection is rare.

Roentgenologic Findings

Multiple small, circular or ovoid, dense spots are situated in many bones, the elongate *lesions lying in the long axis of the bone.* They occur profusely in epiphyses, metaphyses and the small bones of the carpus and the tarsus. Occasional scarce spots may be found in the skull, the ribs and the spine. They are particularly numerous in the pelvis Regions containing cancellous bone seem to be predisposed, whereas the shafts are relatively immune.

Pathology

The trabeculae of the spongiosa are increased in number and thickness.

Differential Diagnosis

Melorheostosis exhibits, in addition to similar small spots, broad bands of dense bone described as blotches which also occupy the shafts of long bones.

11

Diseases of Joints

CLASSIFICATION OF DISEASES AFFECTING JOINTS

1. Infectional arthritis
   - A. Acute (streptococcus, staphylococcus, gonococcus)
   - B. Chronic (tubercle bacillus)
2. Probably infectional
   - A. Rheumatic fever
   - B. Rheumatoid arthritis (atrophic arthritis, proliferative arthritis, chronic infectious arthritis)
   - C. Ankylosing spondylitis (Marie-Strümpell Disease)
   - D. Psoriatic arthritis
3. Toxic arthritis—arthritis associated with various infections
4. Degenerative arthritis (osteoarthritis, hypertrophic arthritis, osteoarthrosis)
   - A. Generalized
   - B. Localized
     - a. Secondary to previous trauma
     - b. Secondary to structural abnormality
     - c. Secondary to rheumatoid arthritis
     - d. Cause unknown
5. Arthritis associated with metabolic diseases
   - A. Gout
   - B. Other metabolic diseases
6. Neuropathic joints
   - A. Tabes dorsalis
   - B. Syringomyelia
7. Neoplasms of joints (cyst, xanthoma, hemangioma, giant cell tumor, synovioma)
8. Traumatic arthritis
   - A. Direct trauma
   - B. Indirect trauma (secondary to postural strain)
9. Systemic disease manifestation
   - A. Serum sickness
   - B. Hemophilia
   - C. Intermittent hydrarthrosis
   - D. Pulmonary osteoarthropathy
   - E. Hysterical joints
10. Local joint disturbances
    - A. Aseptic necrosis
      - a. Known etiology (fracture, dislocation, air embolism)
      - b. Unknown etiology (juvenile osteochondritis or Legg-Calvé-Perthes disease; Kohler's disease; Freiberg's disease; Osgood-Schlatter disease)
    - B. Osteochondritis dissecans (aseptic necrosis?)
    - C. Osteochondromatosis
    - D. Pigmented villonodular synovitis

DEGENERATIVE JOINT DISEASE
(Chondromalacic Arthrosis; Osteoarthritis; Hypertrophic Arthritis; Chronic Osteoarthrosis; Degenerative Arthritis; Arthritis Deformans)

Because this is primarily a degenerative process of the articular cartilage, the term "chondromalacic arthrosis," as suggested by Luck,[1] is most descriptive. The resultant incongruity of joint surfaces produces friction, flaking, and shedding of pieces of cartilage which in turn irritate the synovial membrane. This reactive inflammation of the synovium is the factor which produces pain and is rightfully termed an arthritis which is secondary to chondromalacic arthrosis. This concept explains the alternating periods of exacerbations and remissions.

Etiology

Degenerative changes occur in practically all supportive structures with advancing age. The degree of degeneration varies in different individuals. Cartilage is particularly subject to these changes. The primary lesion (Kellgren,

[1] Luck, J. V.: Bone and Joint Diseases, Springfield, Ill., Thomas, 1950.



FIG. 118 Englemann's disease, microscopic appearance. (*Top*) Cross section of fibula, displaying cortical thickening and regular lamellar structures of the bone. (*Bottom*) Microscopic section (× 40) showing abundant osteoblasts and fibrous marrow structure. Note the absence of mosaic architecture such as occurs in Paget's disease (Griffiths, D. LL : Englemann's disease, J. Bone & Joint Surg. 38B:312)

plaint. A peculiar waddling gait is often noted. With the passage of years, the bony changes extend, weakness becomes pronounced, and pain may become severe. Puberty is late, and the genitalia and secondary sex characteristics are poorly developed.

All laboratory tests are negative.

Roentgenographic Findings

One or several or all of the long bones display a fusiform enlargement of the diaphysis, usually ending abruptly at the metaphysis, but occasionally extending to involve one or both epiphyses The widened dense cortex not only increases the external circumference of the bone but also encroaches upon the medullary canal.

The skull typically is affected by the thickening and sclerosis over the frontal region, the base of the skull at the basiocciput and the petrous portion of the temporal bone.

Diagnosis

Only Caffey's disease (infantile cortical hyperostosis) must be differentiated. The latter is observed within the first year, often before 6 months of age, is invariably accompanied by fever. the increased density is often unilateral, the mandible is frequently affected, and the x-ray changes disappear with complete recovery.

Pathology

Nothing unusual is noted. The cortices are thickened by apposition of mature lamellar bone, both on the periosteal and the endosteal aspects.



FIG. 119. Cartilage in degenerative arthritis. Clefts, Weichselbaum's lacunae, fibrillar appearance and irregular staining qualities are well displayed. (× 108)

lar cortex. New layers of bone thicken the subchondral lamellar plates. The process apparently is in response to increased pressures.

In the intermediate stages, the soft, degenerate cartilage is worn down and completely destroyed This imposes further unusual pressure on the exposed articular cortex which responds by more intensive endochondral ossification. This latter process extends to the joint margins where, in addition to irregular pressures, the stresses imposed by the pull of the capsular attachments are operative. A marginal lip of cartilage forms and is replaced by cancellous bone. The synovial membrane undergoes hyperplasia and villous formation, particularly over fat pads. Inflammatory hyperemia may be present. Reactive synovial changes are less in degree in inactive or nonweight-bearing joints where the factor of irritation is held to a minimum. The synovial membrane may undergo metaplasia at various sites into nests of cartilage cells. These enlarge, become calcified, invaded by blood vessels and replaced by the process of endochondral ossification. The cartilaginous and osteocartilaginous bodies may be extruded into the joint.

In the late stage, the cartilage is completely worn away, and the articular cortex becomes extremely thick, smooth, and polished by the continued friction. Subchondral fractures occur. Subchondral cysts containing amorphous or mucoid material seem to occur at points of greatest stress. Chondromata and osteochondromata continue to develop in the synovial membrane and either remain attached by a pedicle or lie free in the joint. When a joint mouse loses its attachment and therefore its blood supply, its bony core dies while the cartilage covering, which derives its nourishment from synovial fluid, survives and adds further layers of new cartilage. At intervals the surface layer of cartilage calcifies, then becomes covered by a fresh layer of cartilage. The sectioned surface of the joint mouse may display multiple alternating layers of calcified and noncalcified cartilage. Another source of free joint bodies is fracture of marginal osteophytes. The degree of joint degeneration is not proportional to the degree of arteriosclerosis in peripheral or synovial vessels.

Clinical Picture

In spite of the universal normal physiologic aging process and the frequently noted advanced degree of the disease as seen in roentgenograms in many individuals, only about 5 per cent of individuals past 50 have clinical symptoms. The pain is caused by the inflammation in response to joint irritation, whether the latter be caused by pinched synovial villi, mechanical interference by loose bodies, subchondral fractures, or other factors.

1952) is loss of chondroitin, a basic constituent of the matrix. This leaves the collagen fibers without support and fragile to external pressures. The main causes of degeneration beyond the normal physiologic degree, which may be termed "disease," may be classified as follows:

1. **Senescence.** Degeneration normally occurs with advancing age and eventually reaches the severe degree, particularly in weight-bearing joints.

2. **Trauma.** Single or multiple traumata damage and produce further degeneration of cartilage. Such injuries assume a variety of forms, e.g., a direct external blow, pressure of carrying heavy loads, repeated displacements of loose menisci, jagged edges of a fractured fragment, etc.

3. **Static and Shearing Strains.** Malalignment of a joint imposes unequally distributed, increased stresses on one side of a joint. The cartilage succumbs at this point to the abnormal pressure. Examples are: genu valgus or varus, pronated or cavus feet, and an acute lumbosacral angle. Obesity causes both traumatic (increased load) and static (unequal stress) osteoarthritis.

4. **Constitutional.** An unknown factor seems to increase the rate of the degenerative process.

5. **Miscellaneous.** Infection per se does not cause the degenerative process. It destroys the cartilage which in turn becomes irregular and susceptible to frictional forces. Any condition which damages the joint surface or weakens the cartilage, e.g., ochronosis, produces surface incongruities which in turn yield to frictional wear and tear changes and secondary reactive inflammation. The cartilage cannot stand such an environment, being dependent upon healthy synovial membranes and fluid for its nourishment. It deteriorates rapidly. For example, as a result of the damage wrought by rheumatoid arthritis, osteoarthritis frequently supervenes. The relationship of endocrines, particularly the diminution of estrogen at the menopause, is unclear.

Pathology[2, 3]

Normal cartilage is smooth, glistening and bluish white in color. Synovial membranes are smooth and pale and at their attachment to the joint margins they merge with the articular cartilage. Normally, physiologic aging processes start in the second decade and increase with advancing age.[4] Comparable degenerative changes occur in tendons, ligaments, fascia and aponeuroses. Eventually, at the sixth decade thinning and ulcerations of the cartilage and thickening and polishing of the exposed subchondral bone are the usual findings. The following description varies only in degree as an exaggeration of the physiologic process of aging:

Early, degeneration of the hyaline cartilage on the articular surfaces is manifest by a dry, dull, yellowish, opaque and fibrillar appearance. Microscopically, chondrocytes multiply to as many as 20, occupying each enlarged lacunar space (Weichselbaum's lacuna). Later, the cells degenerate and become stellate or amorphous. With friction, the superficial cartilage is shed, and the opened enlarged lacunae cause an irregular pitted surface.

The normal matrix consists of collagenous fiber bundles radially arranged to the surface and a binding substance which occupies the interfibrillary spaces. Both the fibers and the binding substance have the same index of refractility so that the fibers are not visible. When the binding substance degenerates, the fibers become visible, and the decreased resistance of the tissue allows it to split into many clefts. The multiple fissures and fibrillary appearance is characteristic of degenerated cartilage. The superficial tufts of fibrils can be seen grossly when the cartilage is submerged in water. The tiny hairy processes produce a velvety surface. In some areas, the cartilage may proliferate, apparently as an effort to replace the degenerated tissue. However, the proliferated cartilage is undermined by further degeneration and is extruded as a joint mouse. Mucoid degeneration is seen occasionally. In the deeper layers, an attempt at endochondral ossification is manifest by chondrocytes lining up in palisade fashion, calcification of the matrix, and invasion of osteoblastic tissue through openings in the articu-

[2] Luck, J. V.: Bone and Joint Diseases, Springfield, Ill., Thomas, 1950.

[3] Hollander, J. L., *et al.* (eds.): Comroe's Arthritis, Philadelphia, Lea & Febiger, 1953.

[4] Bennett, G. A., Waine, H., and Bauer, W.: Changes in the Knee-Joint at Various Ages, New York, The Commonwealth Fund, 1942.

motion several times daily. This prevents capsular contraction.

3. **Abstain from weight-bearing** on involved joints of the lower extremities. If necessary use crutches.

4. **Reduction of weight** lessens pressure on weight-bearing joints.

5. **Physical therapy**, preferably moist heat followed by massage.

6. **Good body mechanics** by eliminating faulty posture, supporting and exercising the feet, and surgically correcting bony deformities.

7. **Orthopaedic Appliances.** A removable plaster splint secures rest and permits daily physiotherapy. For the back, a simple corset of canvas or plastic suffices. A chair back brace provides comfort for the lower back. An ordinary elastic bandage applied about a joint restricts the extremes of motion and permits a little use. For the entire lower extremity, a long ischial bearing caliper brace removes weight-bearing from the hip, and the addition of a leather cuff about the knee provides immobilization.

8. **Iontophoresis.** Ordinarily, mecholyl or histamine is used. The effect is questionable.

9. **A Warm Dry Climate.**

10. **X-ray Therapy.** This supposedly acts by reducing inflammation and scar-tissue formation.

11. **Hydrocortisone.** A suspension of Compound F injected intra-articularly reduces pain and swelling dramatically within a few hours. Motion is increased. No constitutional effects are noted The steroid acts by its anti-inflammatory action but has no effect upon the degenerative process. Duration of relief of symptoms is variable, lasting from several weeks to many months. A schedule of injections given at regular intervals keeps the patient comfortable and able to continue his activity.

12. **Graduated Exercises.** Weakness of periarticular muscles creates instability and unnatural stresses and strains about the joint, greatly accelerating the destructive process. Strength of the muscles is increased by a program of graded active exercises. Fatigue is avoided.

13. **Drug Therapy.** Thyroid medication is indicated for hypothyroid individuals. It probably acts by weight reduction. Acetylsalicylic acids and salicylates combined with antiacid are specific. These can be combined with an effervescent salt for their psychologic effect. Methyl salicylate rubbed into the skin is said to work by counterirritation, but its beneficial effect is questionable. Vitamin D has been given in massive doses. It causes resorption of calcium from bone, hypercalcemia, metastatic calcifications, renal calculi and renal damage. This author has never observed benefit from its use. Estrogenic hormone may be given to women whose degenerative arthritis dates from the menopausal period. It is particularly effective in relieving pain from Heberden's node. Caution should be exercised in using estrogens. Uncontrollable menorrhagia may result from its use.

14. **Procaine Injections.** A tender spot located about the joint interval may be injected with a local anesthetic. Repeated punctures are made at each sitting. Subsequently, the soreness lasts about 1 day, and the original pain may be relieved dramatically. The mechanism of action is unclear. If improvement is not secured by a second treatment several days later, no further attempts should be made. This indicates that the pathology is more general in distribution and is handled better by intra-articular injection of Compound F.

RHEUMATOID ARTHRITIS

Rheumatoid arthritis is a chronic inflammatory systemic disease of young or middle-aged adults, characterized by destructive and proliferative changes in synovial membrane, periarticular structures, skeletal muscle and perineural sheaths. Eventually, joints are destroyed, ankylosed and deformed.

Etiology

The cause is unknown. Theoretical causes include:

1. **Infection.** Hemolytic and nonhemolytic types of streptococci have been isolated from joints and regional lymph nodes.

2. **Endocrine.** This is suggested by response to adrenocortical steroids.

3. **Allergy.** Rheumatoid arthritics frequently exhibit various allergic manifestations. Eosinophilia is frequent.

4. *Metabolic.*

It is the author's opinion that a toxin lo-



Fig. 120. Osteoarthritis of distal interphalangeal joint. Note bony outgrowths at the margin of base of distal phalanx, clinically termed "Heberden's nodes"

Symptoms

The onset is insidious. A continuous aching *pain* appears, usually mild in degree. It may be localized to one side of the joint or may be generalized about the joint. It is *aggravated by use* and *relieved by rest*. It is *intensified by lowered barometric pressure* which permits of greater synovial swelling. This phenomenon is popularly known as "pain with changes of weather." *Stiffness* occurs with rest and loosens quickly with activity. This symptom is prominent upon arising in the morning. *Heat and salicylates are almost specific* for relief of pain and stiffness

Diagnosis

Findings. The noninflamed joint displays a dry *creaking and grating sensation,* both palpable and audible In the advanced stages with marginal proliferation and capsular thickening, the *joint is enlarged. Motion is limited.* With extreme destruction, barely perceptible motion is present, the joint assuming a fixed deformity, usually in flexion Complete loss of motion never occurs. When the joint is inflamed, an increased amount of synovial fluid is present, and localized tenderness over the joint interval may be noted. No muscle spasm or atrophy is found *Heberden's node* is a characteristic cartilaginous and bony enlargement on the dorsal aspect of the distal interphalangeal joint of a finger. It usually occurs in many fingers. It may occur spontaneously or following trauma. Women are affected most commonly. It may be painless. Or it may appear rapidly with pain, swelling and tenderness. The swelling may be soft or hard. The enlargement is mainly cartilaginous and therefore not visualized in roentgenograms.

Systemic manifestations are absent.

Age. Middle or advanced age.

Sex. Males commonly. When it occurs in women, it generally makes its appearance soon after the menopause.

Joint of Predilection. Terminal interphalangeal joints, lumbar vertebrae, knees, hips, lower cervical vertebrae, sacro-iliacs and elbows.

Patient Type. Frequently overweight.

Laboratory Findings. *Sedimentation rate* normal, blood counts, blood chemistry, and agglutinations with the hemolytic streptococcus are negative. The B.M.R. is decreased in 10 to 30 per cent of patients.

Roentgenographic Findings. Early, the roentgenographic appearance is normal. Then the joint narrowing gradually appears. *Finally,* with progressive degeneration the joint space is narrowed, articular margins sharp, spurring or osteophytic formation at the margins, sclerotic subchondral bone, and bone cysts appear in the subchondral zone at areas of maximum pressure. A negative film does not rule out the disease. *On the other hand, a positive film does* not implicate the joint. One must bear in mind that degenerative changes frequently are superimposed on other disease, notably gout, infectious arthritis and suppurative arthritis.

Treatment

The patient should be reassured and advised that this is not a crippling and deforming type of arthritis. Although degenerative changes are permanent, much can be done to (1) retard progression, (2) alleviate symptoms and (3) provide stability, motion and relief from pain. The following are general nonsurgical measures:

1. **Rest** of the involved joints. This reduces irritation and permits the inflammation to subside.
2. **Move the joint through a full range of**

motion several times daily. This prevents capsular contraction.

3. **Abstain from weight-bearing** on involved joints of the lower extremities. If necessary use crutches.

4. **Reduction of weight** lessens pressure on weight-bearing joints.

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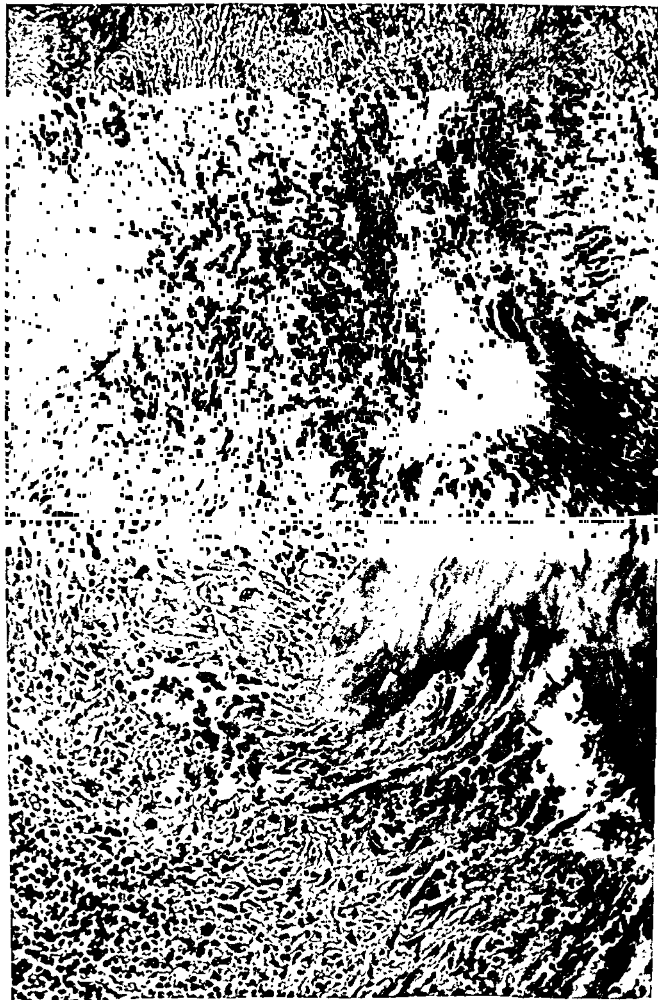


FIGURE 121. (*Caption on facing page*)

calizes in certain tissues which lack an unknown protective factor.

Pathology[5]

The disorder is primarily a synovitis. Early, hyperemia, edema and swelling occur, lining cells proliferate until they are 3 or more layers in thickness, and the underlying tissue is infiltrated with lymphocytes and plasma cells. Villous processes gradually develop and project into the joint cavity. They may become necrotic and extruded into the joint. The typical microscopic lesion is an area of fibrinoid necrosis surrounded by fibroblasts conspicuously arranged radially to the surface of necrosis. Beyond this is an enveloping layer of fibrous tissue. The rheumatoid units and infiltration of round cells are prominent not only in the synovium but also in periarticular structures. The leukocytes frequently are aggregated into round collections which may encircle a blood vessel. An increased amount of clear or slightly turbid fluid accumulates in the joint. The synovium at the periphery forms a pannus of granulation which grows progressively and extends over the articular surface, absorbing and replacing the articular cartilage with fibrous connective tissue. Vascular granulation tissue from the marrow extends toward the articular surface and destroys the cartilage from within the bone. The articular cortex becomes thin and deficient so that fibrous pannus forms the main covering of the bone. The granulation tissue extends toward the opposite articular surface, merging with the pannus there, bridging the joint with granulation tissue. A fibrous ankylosis results. The fibrous tissue may undergo metaplasia into bone. Within the articulating bones, the trabeculae become lessened in number and thinned. The hemopoietic tissue is replaced by a fibrofatty marrow. Fibrous proliferation thickens the capsule.

[5] Herbut, P. A · General Pathology, Philadelphia, Lea & Febiger, 1955.



Fig. 122. Subcutaneous rheumatoid nodules.

Muscle Changes. These are widely distributed in skeletal muscles, the condition being known as "nodular polymyositis." The individual muscle fibers undergo localized or diffuse degenerative changes characterized by increase in size and number of sarcolemmal nuclei, loss of striations, hydropic swelling of fibers, and localized collections of lymphocytes. Similar lesions are formed in the collagen diseases. The degenerated muscle is replaced by fibrous tissue. Loss of muscle elasticity and contractile power is partly responsible for restricted joint motion.

Subcutaneous Nodules. These are composed of the typical basic rheumatoid unit consisting of a central necrotic zone, a surrounding layer of large mononuclear cells radially arranged (palisade formation), and an outer zone of dense connective tissue with marked round-cell infiltration. These nodules are found in 20 per cent of patients, particularly over pressure points as the elbow or over the subcutaneous surface of long bones as over the tibia.

Peripheral Circulation. The arterioles respond poorly to changes of temperature, so that the capillaries are empty or blood flow is sluggish. As a result the distal extremities appear cold and cyanotic.

Lymph Nodes. These exhibit follicular hyperplasia, increased reticuloendothelial activity, proliferation of connective tissue, and lymphocytic invasion of the capsule.

Fig. 121. Rheumatoid arthritis. (*Top*) Microscopic appearance. Showing the area of fibrinoid necrosis surrounded by fibroblasts arranged radially to the surface of necrosis. Round cell aggregations are not prominent in this section. (*Bottom*) High-powered microscopic appearance at the edge of a nodule removed from the olecranon area. Note the radially disposed fibroblasts at the edge of the fibrinoid material, and infiltration of round cells.

Nerves. In the perineural connective tissue are found areas of focal necrosis, epithelioid reaction and leukocytic infiltration.

Cardiac. Rarely, changes identical with those of rheumatic fever are found.

Clinical Picture

The onset is insidious, usually before the age of 40. Females predominate. Constitutional symptoms consist of weakness, fatigue and sweating but no fever. Rarely, the onset may be acute and febrile. Gradually, a number of joints exhibit stiffness and aching, later swelling, pain, warmth, tenderness and limited motion Characteristically, the hands, particularly the proximal joints of the fingers, are involved. In the palm of the hand, the lumbrical muscles are palpable as tender swellings which pull the proximal phalanges into flexion. Next in frequency of occurrence are the feet, the knees and the wrist. Involvement is generally multiple and symmetrical. The swelling of the joint consists not only of increased synovial effusion but also inflammatory edema of the periarticular structures, so that the swelling is maximal about the joint and gradually tapers off at a distance. The overlying skin is stretched, shiny and thin. A typical fusiform appearance results. The joints assume a semiflexed position, and movement in any direction is painful. The surrounding muscles are in spasm. Direction of deformity of the extremities is favored by posture, but certain typical positions are assumed. The arm is in adduction and internal rotation, the elbow in flexion, the forearm in pronation, wrist flexed, and the hand deviated ulnarward; fingers are flexed and deviated ulnarward at the metacarpophalangeal joints, and there is extension at the distal interphalangeal joints. The hip is flexed and adducted, the knee is flexed, and the foot is in equinus, varus and cavus, and the toes are clawed. With persistence of the disease, these deformities become fixed. Sweating, coldness, and cyanosis of the hands and the feet are common. Subcutaneous nodules may be found over bony pressure points or palpable within muscles. The spleen and the lymph nodes may be palpably enlarged.

The course is one of remissions and exacerbations. Remissions occur, characteristically, during pregnancy and jaundice. With each exacerbation, restriction of joint motion becomes progressively worse, a fixed flexion-adduction deformity develops, and muscles which function about the affected joint become atrophic.

Diagnosis

Roentgenologic Findings. Early, the joint structures appear to be normal. Then gradually the articulating bones become osteoporotic and, as the disease progresses and articular cartilage is destroyed, the joint interval is narrowed. The articular cortex becomes thinned and almost indistinct. Finally, bony trabeculations bridge and obliterate the joint space. If the disease stops short of severe destruction, supervening degenerative changes occur consisting of increased density and irregularity of the articular surfaces and marginal spurring.

Laboratory Findings. The erythrocyte sedimentation rate is elevated, particularly during



Fig. 123. Rheumatoid arthritic derelict, a severely advanced stage of the disease.

the active stage. During the periods of remission, the rate continues to increase but to a lesser degree. A hypochromic normocytic anemia is frequently associated. The white cell count is normal.

Serologic Tests.[6, 7, 8] Serum from patients with rheumatoid arthritis contains a substance of unknown composition, the *rheumatoid factor*, which, in the presence of gamma globulin, is capable of agglutinating certain strains of streptococci, sensitized sheep cells, and latex particles. This forms the basis for agglutination tests which are positive in a high percentage of rheumatoid arthritics. The rheumatoid factor may also be present in small amounts in sera of patients with Marie-Strümpell spondylitis, certain collagen diseases and in an occasional normal individual.

A convenient laboratory procedure utilizes a standard suspension of latex particles in a solution of gamma globulin.

1. *Latex Fixation Test on Serum:*
   unknown serum + gamma globulin-latex suspension

Agglutination is likely when the unknown serum contains an abundance of the rheumatoid factor. If no agglutination occurs, the unknown serum contains an inadequate concentration of rheumatoid factor. and a second more sensitive test must be performed.

2. *The Inhibition Test.* This test utilizes the characteristics of euglobulin from the unknown serum. Euglobulin from normal serum will neutralize the rheumatoid factor, thereby inhibiting agglutination. Euglobulin of rheumatoid serum has no effect upon the rheumatoid factor and agglutination proceeds unhindered.

   rheumatoid serum of known high agglutinating activity + unknown euglobulin + standard gamma globulin-latex suspension

This is considered to be the most sensitive test. It is likely to be positive (agglutination occurs) even when the rheumatoid factor is present in minute amounts. When an unknown serum displays a negative latex fixation test and a positive inhibition test, the arthritis in question may be part of a rheumatoid spondylitis or of a lupus erythematosus. The latter disease is of grave import and may be identified by inspecting L-E cell preparations.

Determination of Activity of Disease.[9]

Generalized activity may be measured by the sedimentation rate. The rate during exacerbations increases to as high as 100 mm. per hour and lessens to as low as 30 to 40 mm. It becomes normal only after the disease "burns out." The sedimentation rate as an index is crude, inasmuch as it may be elevated in other conditions including infection, carcinoma, etc.

Localized activity within a single joint is determined by injection of iodized oil into the joint and taking arthrograms. In an acutely inflamed joint the oil is absorbed rapidly and passes to the regional lymph nodes where it is visualized within a few hours to several days. When inflammation is absent and vascularity is reduced, the oil remains unabsorbed for a month or more. It is advisable to perform surgery during periods of minimal activity.

Differential Diagnosis. The main diseases to be differentiated are:

1. *Rheumatic Fever.* As a rule, large joints are involved. After subsidence of arthritis, joints return to normal. Associated with fever, leukocytosis, tonsillitis, cardiac, pulmonary and kidney inflammatory lesions. Younger individuals are afflicted. Responds to salicylates. The antistreptolysin-O titer is elevated (after infection with group A hemolytic streptococci).

2. *Osteoarthritis.* The older age group is predisposed.

Distal IPJ and large weight-bearing joints chiefly

Heberden's nodes—firm fibrous nodule about the distal IPJ

Spurring

No bony ankylosis

No systemic symptoms

ESR not elevated

[6] Hall, A P., Mednis, A. D, and Bayles, T B The latex agglutination and inhibition reactions, New Eng J. M. 258 731, 1958.

[7] Singer, J M, and Plotz, C M Latex fixation test, Am J Med 21:888, 1956

[8] Ziff, M, Brown, P, Baden, J, and McEwen, C.: Hemagglutination test for rheumatoid arthritis with enhanced sensitivity using euglobulin fraction, Bull Rheumat Dis. 5 75, 1954

[9] Kelikian, H.: Surgery in the treatment of chronic arthritis, S. Clin North America 29:87, 1949.

Treatment

Rheumatoid arthritis is an inflammatory disease destructive to joints. Involvement of one joint leads to secondary changes detrimental to other joints in the extremity. Therefore, the aim of treatment is to keep the inflammatory process at a minimum, thereby preserving joint motion, maintaining health of muscles supplying motor power about the joint and preventing secondary joint stiffness and deformity. In addition, constitutional defects must be corrected, notably the secondary anemia. The possible deformities must be anticipated and prevented by appropriate splinting. Finally, surgical measures correct the deformities, eliminate pain and provide stability. A plan of treatment should be outlined and followed through.

Conservative Treatment.[10]

Rest. The patient is kept at complete bedrest.

Removal of Foci. Teeth, tonsils, sinuses and pelvic organs are investigated, and infections are eliminated.

Nutritious Diet. A high caloric, high vitamin diet.

Transfusions and Hematinics. These are continually necessary during the entire course of the disease.

Hormones. Combinations of estrogen and androgen are administered for their anabolic effect on bone structure.

Dilute Hydrochloric Acid. This is necessary to combat the achlorhydria which contributes to the anemia.

Splinting. The inflamed joint is firmly immobilized in a plaster splint. This relieves pain and reduces inflammation quickly.

The position of function is desirable in the event that ankylosis ensues. Several times daily the cast is removed, hot packs are applied, or the patient is placed in a Hubbard tank at 92.6° to 102° F., and the joints are put through a full range of motion. This helps to maintain joint mobility. While immobilized, muscle-setting exercises combat muscle weakness and atrophy. After removal of the splint resistance exercises are prescribed to restore joint stability before allowing weight-bearing. Recent experience has shown that this treatment reduces the incidence of ankylosis.[11,12]

If weight-bearing is permitted on an unstable joint, the instability causes recurrent effusion, muscle-wasting and further instability, a vicious cycle.

Positions of Rest for Inflamed Joints:

Shoulder—scapulohumeral angle 45°; forward flexion 45°

Elbow—70° flexion, 15° supination of forearm

Wrist—30° dorsiflexion, support to arches of hand

Fingers—45° flexion

Spine—full extension, good posture

Jaw—open at least 1 inch

Hip—flexion 5°, abduction 5°, neutral rotation

Knee—flexion 5°

Ankle—at right angle, no valgus or varus of foot

Cortisone. In 1935 Kendall at the Mayo Clinic isolated Compound E (17 hydroxy-11-dehydrocorticosterone) from the adrenal cortex. In 1949 Hench et al. discovered the therapeutic effect of Compound E or cortisone. Later, he noted the effect of pituitary adrenocorticotropic hormone (ACTH) which stimulates the adrenal cortex to secrete a cortisone-like substance, probably Compound F (17-hydroxycorticosterone). The circumstances leading to the discovery was the observation that there was a remission of rheumatoid arthritis during pregnancy and jaundice, and the adrenal cortex underwent hypertrophy and hyperfunction in pregnancy. Compounds E and F produce a dramatic reduction of inflammation and remission in rheumatoid arthritis. The acute inflammatory swelling of the joints is reduced, and mobility is restored. Unfortunately, cessation of treatment is usually followed by relapse, and continued treatment may cause complications. Cortisone is useful in controlling the disease. It has no curative effect. Its benefit is to reduce inflammation, prevent further destruction, maintain mobility, and avoid muscle atrophy and secondary effects to neighboring joints. One hopes

[10] Kuhns, J. G.: Non-surgical treatment of arthritis, Instructional Course Lectures, Am. Acad. Orth. Surg. 6.292, 1949.

[illegible]

diseases, J. Bone & Joint Surg. 34B:211, 1952.

to maintain the integrity of the joints and to prevent deformity until the disease has run its course. The commonly accepted duration is 3 to 5 years.

Effect of Cortisone on Histopathologic Lesions.[13] The synovial membrane tends to revert to normal. The lymphocytes and the plasma cells disappear, vascularity is decreased, and edema subsides. The subcutaneous nodules become smaller and less inflammatory. The central necrotic zone becomes poorly defined, the palisade layer disappears, the outer zone of fibrous tissue becomes more dense, and round cells are sparse. In muscle, the nodules disappear, and degenerative changes are less noticeable.

Method of Administration. Cortisone is effective when given orally. Beneficial effects may be seen within 2 to 3 hours and persist for 6 to 12 hours. The starting dose is 100 mg. per day in 3 or 4 divided doses, and this is increased if necessary or reduced to a minimum maintenance dose. The majority of patients are controlled on about 50 mg. per day. The treatment is preferably initiated in a hospital where the effects can be measured by laboratory procedures. Adverse findings call for reduction of dosage or complete stoppage. It is desirable to limit each course of treatment to a period of a few months with rest periods of several weeks between courses. When discontinuing medication, symptoms of depression and weakness appear which are readily controlled by small doses of Dexedrine (amphetamine sulfate).

If cortisone is administered with para-aminobenzoic acid, its effect is markedly enhanced. Therefore, doses as low as 25 mg. may be effective.

Systemic Effects of Cortisone:

1. Depresses pituitary ACTH secretion
2. Causes adrenal cortical atrophy (reversible). Decreased excretion of 17-ketosteroids
3. Depression of adrenal cortical secretion
4. Inhibition of Koch phenomenon
5. Reduces antibody production.
6. Diabetogenic: increased glucogenesis, hyperglycemia, glycosuria, insulin resistance, liver glycogen increase. Cortisone increases the insulin requirement in diabetics.
7. Catabolic: Nitrogen-containing tissue destruction shown by negative nitrogen bal-

[13] Norcross, B M., Lockie, L. M., Constantine, A. G., Talbott, J. H., and Stein, R. H.: The effect of cortisone and ACTH on the histopathologic lesions of rheumatoid arthritis, Ann. Int. Med. 36.751, 1952.



FIG. 124. Pathologic fractures of 1st and 4th lumbar vertebrae in the course of cortisone therapy. Note that these are not wedge-shaped compressions. The fractures have occurred centrally from expansile force of the disks, a pertinent point establishing this as a pathologic type of fracture. Until fracture occurs, the vertebral bodies of the rheumatoid spine, in spite of being osteoporotic, tend to retain their size and configuration.

TREATMENT

Rheumatoid arthritis is an inflammatory disease destructive to joints. Involvement of one joint leads to secondary changes detrimental to other joints in the extremity. Therefore, the aim of treatment is to keep the inflammatory process at a minimum, thereby preserving joint motion, maintaining health of muscles supplying motor power about the joint and preventing secondary joint stiffness and deformity. In addition, constitutional defects must be corrected, notably the secondary anemia. The possible deformities must be anticipated and prevented by appropriate splinting. Finally, surgical measures correct the deformities, eliminate pain and provide stability. A plan of treatment should be outlined and followed through.

Conservative Treatment.[10]

REST. The patient is kept at complete bedrest.

REMOVAL OF FOCI. Teeth, tonsils, sinuses and pelvic organs are investigated, and infections are eliminated.

NUTRITIOUS DIET. A high caloric, high vitamin diet.

TRANSFUSIONS AND HEMATINICS. These are continually necessary during the entire course of the disease.

HORMONES. Combinations of estrogen and androgen are administered for their anabolic effect on bone structure.

DILUTE HYDROCHLORIC ACID. This is necessary to combat the achlorhydria which contributes to the anemia.

SPLINTING. The inflamed joint is firmly immobilized in a plaster splint. This relieves pain and reduces inflammation quickly.

The position of function is desirable in the event that ankylosis ensues. Several times daily the cast is removed, hot packs are applied, or the patient is placed in a Hubbard tank at 92.6° to 102° F., and the joints are put through a full range of motion. This helps to maintain joint mobility. While immobilized, muscle-setting exercises combat muscle weakness and atrophy. After removal of the splint resistance exercises are prescribed to restore joint stability before allowing weight-bearing. Recent experience has shown that this treatment reduces the incidence of ankylosis.[11, 12]

If weight-bearing is permitted on an unstable joint, the instability causes recurrent effusion, muscle-wasting and further instability, a vicious cycle.

POSITIONS OF REST FOR INFLAMED JOINTS:

Shoulder—scapulohumeral angle 45°; forward flexion 45°

Elbow—70° flexion, 15° supination of forearm

Wrist—30° dorsiflexion, support to arches of hand

Fingers—45° flexion

Spine—full extension, good posture

Jaw—open at least 1 inch

Hip—flexion 5°, abduction 5°, neutral rotation

Knee—flexion 5°

Ankle—at right angle, no valgus or varus of foot

CORTISONE. In 1935 Kendall at the Mayo Clinic isolated Compound E (17 hydroxy-11-dehydrocorticosterone) from the adrenal cortex. In 1949 Hench et al. discovered the therapeutic effect of Compound E or cortisone. Later, he noted the effect of pituitary adrenocorticotropic hormone (ACTH) which stimulates the adrenal cortex to secrete a cortisone-like substance, probably Compound F (17-hydroxycorticosterone). The circumstances leading to the discovery was the observation that there was a remission of rheumatoid arthritis during pregnancy and jaundice, and the adrenal cortex underwent hypertrophy and hyperfunction in pregnancy. Compounds E and F produce a dramatic reduction of inflammation and remission in rheumatoid arthritis. The acute inflammatory swelling of the joints is reduced, and mobility is restored. Unfortunately, cessation of treatment is usually followed by relapse, and continued treatment may cause complications. Cortisone is useful in controlling the disease. It has no curative effect. Its benefit is to reduce inflammation, prevent further destruction, maintain mobility, and avoid muscle atrophy and secondary effects to neighboring joints. One hopes

[10] Kuhns, J. G : Non-surgical treatment of arthritis, Instructional Course Lectures, Am Acad. Orth Surg 6 292, 1949.

[11] Duthrie, J. R.: Discussion during symposium on management of rheumatoid arthritis, British Orthopedic Association Annual Meeting, 1951.

[12] Duthrie, J. R : Medical treatment of rheumatic diseases, J. Bone & Joint Surg. 34B:211, 1952.

peated weekly over 4 to 5 months until a total of 1 Gm. has been given. A favorable response is indicated by clinical improvement and reduction of the E.S.R. With each dose, 10 cc. of 10 per cent calcium gluconate is given to lessen possibility of reaction. Another course of treatment is given after 6 to 12 months or after a recurrence. If no improvement occurs after 5 months, it is useless to continue. Change of symptoms should be expected between 6 and 15 weeks. Recurrence can occur from 6 months to 5 years.

Toxic Reactions. Eternal vigilance is mandatory to prevent serious toxic reactions. Before each injection the skin is examined for a rash or purpuric spots. Pruritus is a complaint before appearance of the rash. The mouth is examined for stomatitis. The patient is questioned for gastro-intestinal symptoms, especially diarrhea. The urine is checked for albumin or microscopic hematuria. A slight amount of albuminuria is permissible. Hematologic reactions include granulocytopenia, thrombocytopenia and aplastic anemia. These are rare but serious. They are less likely with the small dosage schedule herewith recommended. Every 2 weeks a complete blood count and platelet count is done. The reduction of hemoglobin, leukocytes and platelets and the tendency to eosinophilia is a danger signal, and all treatment must be suspended. If improvement does not occur promptly after discontinuance of treatment, British Anti-Lewisite (BAL) is administered without delay. From 2 to 3 mg./Kg body weight of 10 per cent BAL is injected deeply intramuscularly—6 times the first day, 4 times the second and the third days, then 3 times daily for 4 days.

Contraindications. Hepatic or renal damage, blood dyscrasias.

COMBINED CORTISONE AND GOLD THERAPY. Such treatment seemingly would be advantageous, but the percentage of patients improved is only a little greater than when gold is employed alone. The excellent therapeutic response may continue for a long time after the cortisone has been discontinued.[18]

NITROGEN MUSTARD: ITS EXPERIMENTAL USE.[19] A systemic cytotoxic effect of mustard gas and related compounds is characteristic of nitrogen and sulfur derivatives; it affects chiefly the proliferative tissues, especially those of lymphoid nature. The administration to rats causes, like ACTH, an appreciable decrease in the adrenal ester cholesterol and a mild decrease in ascorbic acid promptly. Toxic manifestations occur later. The effect may be due to action directly on the adrenal cortex or on the pituitary gland influencing the production of ACTH. In the few clinical cases on which it was tried, prompt but transient improvement was noted. Its use is not recommended.

MANIPULATION. Passive manipulations or wedging casts to overcome contractures is not advisable. It is impossible to estimate the extent of intra-articular adhesions. These adhesions may be torn only to be replaced by more extensive scarring.

The fibrotic inelastic muscles may be damaged. Fracture of the osteoporotic bones is a definite risk. Where the articular involvement is considered minimal, gentle prolonged traction or, as in the deformities of the hand, dynamic splinting is permissible. Otherwise, gradual spontaneous correction by active motion is most desirable and the result more lasting. Deformities which are persistent should be treated surgically.

X-RAY TREATMENT. This is used occasionally for relieving pain and low-grade inflammation, particularly where cortisone, ACTH and gold salts have been ineffective. Its greatest field of usefulness is in rheumatoid arthritis of the spine.

RELIEF OF MUSCLE SPASM. Hot, wet packs are most beneficial. Muscle relaxants as myanesin (Tolserol) and prostigmin are of doubtful value.

ASPIRATION. Removal of joint fluid is indicated when effusion is recurrent and causes pain by distention.

HYDROCORTONE INJECTIONS. When only one or two large joints are involved, compound F injected intra-articularly is effective in rapidly reducing the inflammation and eliminating pain. The local effect is prolonged,

[18] Thompson, H. E.: Cortisone and gold therapy in chronic rheumatoid arthritis, Ann Int. Med. 36 992, 1952.

[19] Shutkin, N. M : Note on the use of nitrogen mustard in rheumatoid arthritis, J. Bone & Joint Surg. 33A:265, 1951.

ance. This delays development of new bone The bony trabeculae becomes thinned and the general architecture weakened, making bones susceptible to trauma. Compression fractures of the spine are frequent.

8. Electrolyte balance effect: Serum potassium level falls, cardiac arrhythmias result Sodium retention occurs early, followed later by *increased excretion*. Early edema causes gain in weight, possible hypertension Hypochloremic, hypokalemic alkalosis develops.
9. Involution of lymphatic tissue
10. Delays development of granulation tissue.
11. Drop in circulating eosinophils
12 Cushing syndrome features—cutaneous striae, moon facies
13. Acute psychosis: euphoria, stimulation, insomnia
14. Increased appetite

Before treatment one must determine the presence of tuberculosis and diabetes, both of which may flare up under the influence of cortisone. Other contraindications are chronic nephritis, acute psychosis and peptic ulcer. During treatment, a low-sodium diet is prescribed. Increased blood pressure, increased weight and pitting edema are danger signals, indicating sodium retention. Blood sugar, sodium and potassium and blood counts are checked at intervals The erythrocyte sedimentation rate may be done several times a year to record activity of the disease. After treatment with cortisone, one must exercise caution regarding surgical procedures. The adrenal cortex may be atrophied and unable to respond to the stress of surgery. Small doses should be prescribed, both preoperatively and postoperatively.

ADRENOCORTICOTROPIC HORMONE (ACTH). The hormone from the pituitary stimulates the adrenal cortex to produce more Compound E and F, the effects of the latter being reproduced. The adrenal cortex is hypertrophied, and there is an increase of urinary 17-ketosteroids. The hormone is effective only by parenteral administration. It is prepared in a gelatin menstruum which retards its absorption from the site of injection and prolongs its action. It is injected in 100 mg. daily doses. The usual precautions are taken.

SALICYLATES. Although not having its seemingly specific effect for rheumatic fever, it is used chiefly for analgesia. However, salicylates have an action comparable with corticotropin and/or cortisone when given in large doses. It has been shown experimentally that salicylates work through the pituitary as the mediator, as proved by loss of its effect after hypophysectomy. These effects include: eosinopenia, *inhibition of inflammatory phenomena* and tissue sensitivity, depression of gamma globulin and antibody production, retention of sodium chloride and water, loss of potassium and nitrogen, decreased glucose tolerance, increased excretion of uric acid, and increased 17-ketosteroids in the urine; also, the depletion of adrenal cholesterol and ascorbic acid and even occasional production of a Cushing's syndrome.[14]

GOLD THERAPY.[15] Gold salts when used judiciously have a prolonged beneficial effect. In contrast, prolonged benefit from ACTH and cortisone is unusual. The mechanism of action of gold is unknown; its use is empiric. However, experimental use of the radioisotope of gold reveals selective uptake in synovia and other articular tissues as well as liver, kidney and spleen.[16] However, in rheumatoid arthritis, the affinity for the synovial tissues is strong. It may also work by altering the immune response because of its affinity for reticuloendothelial tissue.[17] During treatment 75 per cent is retained in the tissues, while 25 per cent is excreted. The amount in the body increases during treatment, and excretion continues for many months after discontinuing treatment. Remission occurs only while gold is in the body and is being excreted. Gold is of value only during the acute stage. The best available preparations at the present time are Myochrysine and Solganol-B.

Procedure (with Myochrysine). Injections are given deeply intramuscularly. The first injection is 10 mg.; 1 week later, 25 mg.; the third week, 50 mg. The 50-mg. dose is re-

[14] Pfizer Spectrum, *in* J.A.M.A., Vol. 151 (No 9 Adv), February 28, 1953

[15] Hersperger, W. G : Gold therapy for R.A , Ann Int Med. 36 571, 1952.

[16] Bertand, J. J., Waine, H , and Tobias, C. A : Distribution of gold in the animal body in relation to arthritis, J. Lab & Clin Med. 33:133, 1948.

[17] Kersley, G D.: The Present Status of Gold Therapy in R. A , Practitioner 161 158, 1948.

phalangeal joints and extends at the interphalangeal joints. If the proximal interphalangeal joint is hyperextended, the distal joint is flexed. Contracture of the adductor muscles draws the thumb into the palm toward the third metacarpal. This is the "intrinsic plus" position. The collateral ligaments at the metacarpophalangeal joints are so degenerate that the proximal phalanges are subluxated far anteriorly and fixed there by ligament and muscle contracture. The fingers deviate ulnarward at the metacarpophalangeal joints as the long extensors are displaced into the interknuckle grooves. The thumb is overflexed by the short flexors at the metacarpophalangeal joint, the long extensors hyperextending the distal joint.

Occasionally, the long extensors predominate. The fingers are hyperextended at the metacarpophalangeal joints, flexed at the middle joints and extended at the distal joints.

In the early stage of muscle irritation and spasm, a reverse knuckle bender splint and a spring cockup splint will apply mild elastic traction counteracting deformity.

After the process has become quiescent, correction of muscle balance and release of muscle tension are aims in surgical treatment. The following procedures are commonly used:

1. *Tenotomy of lateral bands* allows metacarpophalangeal joints to extend and distal two joints to flex.

2. *Shortening of metacarpals* releases tension on all muscles. A segment of bone is removed at the base and fixed temporarily with Kirschner wires. (This procedure is done only when metacarpophalangeal joints can be extended passively.)

3. *Arthroplasty*. Excise metacarpal heads. (Procedure is done only when metacarpophalangeal joints cannot be passively reduced.) The new joints should be pinned in alignment until enough fibrous tissue contracts around them to provide stability.

4. *Shortening of the thumb metacarpal* at the base. Fibrotic adductors may be removed and the cleft kept open by pins and subsequent pedicle graft. If adduction is lost, a tendon-T operation is done. Tendon transference may be necessary to restore opposition.

5. *Radial displacement of extensor tendons* to overcome ulnar drift. The luxated tendons are elevated, transferred to the radial side of the knuckle where it is sutured into a slit in the dorsal aponeurosis.

6. *Tendon transfer*. The extensor indicis proprius and extensor digiti quinti proprius are transferred to the lateral band on the radial side of the index and the little fingers respectively.

Hips. Some type of arthroplasty or pseudarthrosis is usually indicated because as a rule both hips are involved. Cortisone does not seem to affect the final result.

Arthroplasty of Smith-Petersen (1939). A vitallium mold is interposed, the articular surfaces become lined with smooth, glistening fibrocartilage, and the underlying bone becomes more firm. Reankylosis is less likely than with other substances as fascia lata. Recurrence of stiffness and deformity is greater in rheumatoid than in degenerative arthritis, but some useful motion is gained, permitting ambulation with canes and sitting on a high chair.

Technical principles:

1. Acetabulum is made as large as possible yet consistent with stability of the mold and the femoral head.

2. Joint capsule and synovial membrane are excised completely, together with rheumatoid pannus.

3. Blood transfusions are necessary because of considerable hemorrhage.

4. Coagulate raw bone surfaces after reshaping; this prevents new bone formation.

5. Modification adapted to rheumatoid arthritis to gain more mobility in spite of marked fibrosis:

A. *Whitman reconstruction.* Remove head, *reshape neck,* displace greater trochanter downward before placing the Vitallium cup.

B. *Colonna.* Remove head and neck, *reshape trochanter* to form new articulating surface. Trochanteric muscles are detached and replaced lower down.

Arthrodesis. Bilateral arthrodesis is not advisable. However, with one freely movable hip joint, the other can be fused, providing stability and freedom from pain.

Prosthetic Replacement. Devices for replacing the femoral head are not advised. The

and constitutional effects are avoided. One cc. of the suspension is injected, and the patient usually experiences some increased discomfort for about 1 day. Thereafter, improvement is dramatic. The injection is repeated at weekly intervals, then reduced in frequency until the maintenance dose and length of remission are determined.

Surgical Treatment.[20, 21, 22] Treatment by surgery should not be delayed too long. Smith-Petersen showed that a primarily involved joint, if left deformed and mechanically deficient, will cause secondary changes in other joints, and severe, fixed deformities will ensue. Delay until the disease is "burnt out" increases the difficulty. The muscles become fibrous, ligaments and capsules become masses of scar tissue, and secondary joints undergo degenerative changes. However, one may avoid surgery during a period of activity when constitutional effects are at their height. The period of decline of the ESR is strategic.

Cortisone administered over a period of time induces atrophy and hypofunction of the adrenal cortex. Such a patient is highly susceptible to the shock of surgical trauma. Preoperative preparation demands discontinuing cortisone and giving, instead, daily large doses of ACTH for several weeks until the adrenal cortex has been restored. Transfusions and liver-iron preparations are given to combat anemia.

OPERATIONS IN EARLY DISEASE

Shoulder: *Excision of the Acromion* (Smith-Petersen). This relieves pain of a subdeltoid bursitis, frees the tendinous cuff and allows excision of a congested villous bursa. Relief of pain permits gradual restoration of motion by active exercise.

Section of adductors and subscapularis to correct adduction contracture.

Rotation osteotomy of humerus to overcome internal rotation contracture.

Arthrodesis, if involvement is unilateral and severe.

Elbow: *Excision of the Radial Head* Spasm of the biceps, a prominent feature of the early stages, draws the head upward against the capitellum, damages the joint surfaces and causes pain and further muscle spasm. Synovectomy is done at the same time. The procedure relieves pain quickly and permits free motion.

Knee: *Synovectomy.* This is done for recurrent effusion with synovial thickening. If the articular cartilage is eroded extensively, synovectomy alone is useless and may result in an unsound fibrous ankylosis.

Tenotomies

Posterior Capsulotomy (Wilson)

INTERMEDIATE AND LATE OPERATIONS

Elbow—*Arthroplasty.* Excision should be thorough to prevent reankylosis. Early, considerable instability is counteracted later by periarticular fibrosis. Motion is good but not stable enough for crutch walking.

Arthrodesis. When fixed in the functional position, a painless strong limb is provided for heavy activity.

Wrist. *Radiocarpal fusion in the "Grasp Position."* The wrist is fixed in 10° to 15° of dorsiflexion. If flexion deformities of the fingers due to fibrotic flexors and intrinsics are present, dorsiflexion at the wrist will increase the degree of flexion, making it more difficult to open the hand. Therefore, surgical correction of the hand should logically follow. Wrist fusion should be carried out early, before deformity is extreme and before there is gross destruction at the metacarpophalangeal and the interphalangeal joints

Resection of Lower End of Ulna. The indication is limited supination and pronation due to rheumatoid involvement of the distal radioulnar joint.

Hand.[23] Degenerative changes in the joints make fingers easy prey to deforming contractions of muscles acting about the joints. Contracture of these muscles, particularly the intrinsics, results in muscle imbalance between the long extensors, the long flexors and the intrinsics (A similar mechanism occurs in ischemic contracture local in the hand, collagen diseases and various types of arthritis) Intrinsic contracture flexes the metacarpo-

[20] Law, W A Surgical treatment of the rheumatic diseases, J. Bone & Joint Surg 34B 215, 1952

[21] Speed, J S. Campbell's Operative Orthopedics, St. Louis, Mosby, 1949

[22] Badgley, C. E The orthopedic treatment of arthritis, Am. Acad Orthop Surgeons, Lect 5 314, 1948

[23] Bunnell, S.: Surgery of the rheumatic hand, J. Bone & Joint Surg 37A.759, 1955

TABLE 3. CLINICAL DIFFERENTIATION

	RHEUMATOID ARTHRITIS	OSTEOARTHRITIS
Geographic distribution	Most common in temperate climates; rare in the tropics	Climate not a factor
Family History	Often a history of rheumatic fever or rheumatoid arthritis in an immediate member of family	Frequently, a history of a similar form of arthritis in one or both parents
Past History	Occasionally, a history of rheumatic fever; frequently, of tonsillitis or sinusitis	Not characteristic; sometimes history of trauma or faulty body mechanics.
Age at onset	Any age; over 80 per cent between 20 and 50	Rare before 40
Mode of onset	Rarely acute; usually subacute or insidious; often accompanied by migratory pains	Insidious; not accompanied by migratory pains
Patient's general condition	Usually undernourished, anemic, and "chronically" ill; frequently slight fever (+ 99° F.) and slight leukocytosis	Well nourished, frequently obese; not anemic; no fever, no leukocytosis
Involvement of joints	Symmetrical and generalized; proximal interphalangeal joints especially involved	Usually weight-bearing joints, spine, hips, knees; distal joints of fingers (Heberden's nodes)
Appearance of joints	Early: periarticular swelling, fusiform fingers Late: ankylosis, extreme deformity, ulnar deflection	Early: slight articular enlargement Late: more pronounced articular enlargement; limitation of motion usually slight; never ankylosis; Heberden's nodes
Muscular atrophy	Often pronounced, particularly in later stages	Not characteristic
Cutaneous changes	(1) Extremities frequently cold and clammy; skin atrophic and glossy; redness of thenar and hypothenar eminences (2) Psoriasis occasionally present	No characteristic features
Subcutaneous nodules	Present in 15 to 20 per cent of cases	Not present

TABLE 4. LABORATORY DIFFERENTIATION

	RHEUMATOID ARTHRITIS	OSTEOARTHRITIS
Agglutination reactions	Positive in over 50 per cent of typical cases	Never definitely positive
Sedimentation rate	Usually greatly increased; tends to return to normal as patient improves	Normal or only slightly increased
Roentgenologic appearances	Early: osteoporosis, periarticular swelling and joint effusion Late: narrowing of joint space, bone destruction, ankylosis and deformities	Early: no osteoporosis; slight lipping at joint margins Late: marked lipping, osteophytes, narrowing of joint space, deformation of articular bone ends.

bony structure is inadequate for holding the prosthetic device securely in place.

Pseudarthrosis. The procedures of Girdlestone (1947) and Batchelor (1948) restores movement and corrects deformity. Instability is unlikely because of surrounding soft-tissue fibrosis.

Knee. *Arthrodesis* is most effective, providing a painless stable limb.

Arthroplasty is unsatisfactory. Instability and reankylosis are frequent.

Technic. Complete exposure is accomplished through a long anteromedial incision. The synovium, the fat pad, the menisci, the pannus and the fibrillated cartilage are removed. The synovial membrane may be left in the posterior compartment. Postoperatively, the limb is suspended, and active and passive motion, baking and massage are given. Subsequently, a knee brace with control dial is worn.

Osteotomy. Supracondylar section overcomes moderate flexion deformity. Advised only when some painless motion is present. Capsulotomy and tenotomies are of no value.

Ankle Joint. Equinus deformity is corrected by:

Achilles tendon lengthening and posterior capsulotomy.

Panastragaloid arthrodesis if joint involvement is severe.

Feet and Toes. The foot presents a rigid clawed and cavus deformity. For hammer toe, resection of the proximal phalanx or interphalangeal arthrodesis is done. The heads of the second, the third and the fourth metatarsals are excised. For hallux valgus, a Keller procedure is advisable A triple arthrodesis corrects the cavus deformity.

Spine. *Osteotomy* corrects the forward stooped deformity at the level of L2 or L3 A V-shaped excision of bone is performed across the line of the articular processes, from interlaminar space to the intervertebral foraminae Then the deformity is corrected by hyperextension. This is followed by spine fusion and plaster immobilization. (See Marie-Strümpell disease.)

Miscellaneous Procedures: Lumbar sympathectomy[24] is done in the vasospastic types. It improves peripheral circulation and relieves pain.

[24] Adson, A W., and Rowntree, L. G The surgical indications for sympathetic ganglionectomy and trunk resection in the treatment of chronic arthritis, Surg. Gynec. & Obst. 50.204, 1930

STILL'S DISEASE
(Rheumatoid Arthritis in Children)

Rheumatoid arthritis in the infant or the child tends to be more acute than in the adult. It is characterized by fever, swelling of many joints, enlargement of the spleen and the lymph nodes, and rapid development of fixed flexion deformities, muscle wasting and ankylosis. The most frequently involved joints are the knees, the wrists and the cervical spine. The enlarged hard nontender lymph nodes are the regional nodes of the involved joints.



Fig 125. Still's disease or rheumatoid arthritis in a child.

active sclerosis, large exostoses, multiple loose intra-articular bodies, deformity, subluxation and dislocation, and marked parosteal ossification. When destruction and osteoporosis predominate, the *atrophic or degenerative type* is present. The *hypertrophic or proliferative type* is characterized by excess bone formation in the form of extremely dense sclerosis, large osteophytes, and parosteal ossification. Usually, both types are present in the same joint.

Grossly, the capsule is thickened and hyperplastic. It is stretched by repeated effusions of large amounts of synovial fluid until it becomes redundant. Its attachments about the joint margins become progressively displaced distally as the articulating bone ends are destroyed. The articular aspect of the capsule is lined by a ragged synovial membrane from which villi grow inward, particularly at the joint line. The membrane contains cartilaginous and bony placques some of which are partially extruded remaining attached by a pedicle while others are completely separated and lie free within the joint cavity. The ligaments are stretched, permitting unusual mobility of one bone upon the other. The articular cartilage is degenerate and worn away. The exposed subchondral bone is necrotic, fractured and compressed or eroded. The underlying cancellous bone may be porous and filled with debris. More commonly, reactive sclerosis forms dense bone which replaces the cancellous bone. A pannus of granulation tissue may extend over and absorb the articular cartilage. Marginal exostoses form by reactivated endochondral ossification and are usually massive. The joint cavity is filled with debris of pieces of necrotic bone and cartilage. In the neighboring muscles and fascia, heterotopic bone formation takes place.

Microscopically, the capsule exhibits fibroblastic proliferation, organizing hemorrhage due to tears, and bone formation by metaplasia or preceded by cartilage formation. The articular cartilage displays typical degeneration, i.e., loss of matrix, fibrillations and fissuring. Where it is not worn away by bony contact, it is eroded by a pannus. Within the bone, multiple small fractures and areas of necrotic bone are seen. In some areas osteoclastic resorption of dead bone takes place. In the immediate vicinity active bone formation replaces the cancellous trabeculae with dense laminated compact bone. When bone replacement is deficient, the marrow spaces become filled with amorphous debris and subject to erosion.

Clinical Picture

Trauma with or without fracture frequently initiates the condition. A large amount of synovial fluid greatly distends the joint, and the overlying tissues appear edematous. The fluid when aspirated is abundant, yellow, viscous, and clots rapidly. The cell count is 500 to 2,000 per cu. mm., mainly lymphocytes. Gradually, the swelling subsides leaving a relaxed capsule and an abnormally mobile joint. For example, the knee will display lateral mobility or can be hyperextended. Over the ensuing weeks or months the joint becomes enlarged and deformed and even more unstable. Repeated joint effusions occur. Pain is notably absent. Examination of the joint reveals marked irregularities identified as bony projections from the articulating bones and bone formations in the surrounding soft tissues. Palpation of the redundant soft thickened capsule reveals many intra-articular bodies similar to a "bag of bones." The joint can be passively and painlessly moved in all directions.

In tabes dorsalis, the lower extremities and the spine are prone to involvement. Associated signs of tabes include ataxia, Argyll Robertson pupils, absent knee reflexes and absent deep position, vibration and pain sense. Symptoms complained of are lancinating pains, girdle pains, paresthesias, gastric crises and loss of bladder control.

In syringomyelia, glial proliferation and cavitation occur about the central canal of the cord in the lower cervical and upper dorsal region. Therefore, the arthropathy is confined mainly to the upper extremities. Clinically, one finds sensory dissociation—loss of pain and temperature and preservation of touch. Deep sensation is undisturbed. Progressive muscle atrophy in the arms, and fibrillations and trophic changes in the fingers are added findings. Most commonly, the elbow is involved. It is swollen with excess fluid, destroyed, displays abnormal lateral mobility, and bony masses in the soft tissues are greater than in tabes. The cervical spine when involved causes a kyphosis or scoliosis, but the cord is not involved.

Endocardial, pericardial, pleural and nephritic lesions are often present. Dimineralization of the bones and narrowing and obliteration of the joints are noted in roentgenograms. Maturation of the bones about the inflamed joints is accelerated. This results in brachydactylia. Regional external cortical thickening occurs in the small tubular bones of the hands and the feet. Laboratory findings include leukocytosis, anemia and an elevated sedimentation rate. Remissions and exacerbations over 6 or 7 years are the rule.

Treatment consists of complete bedrest, nutritional care, and immobilization of the affected joints to reduce inflammation and to prevent contractures. If the child will cooperate, active muscle setting exercises are done. Once daily the plaster splints are removed, hot packs are applied, and the joints are put through a full range of motion. Gold salts, cortisone and ACTH dramatically relieve pain, reduce swelling and permit mobilization of joints. These measures control the disease until it "burns out." Residual deformities require surgical intervention.

PALINDROMIC RHEUMATISM[25, 26]

Palindromic rheumatism is a rare, benign condition characterized by multiple recurring attacks of painful inflammation affecting joints and adjacent tissues. The cause is unknown. Each attack lasts but a few hours to a day or two and is followed by a complete remission. All joints are liable to involvement, but the finger joints are predisposed. The typical attack begins very suddenly, usually late in the afternoon Within a few minutes a joint may become painfully swollen, reaching its intensity within a few hours. The periarticular soft tissues are reddened and swollen, the overlying skin stretched and shiny. Disability is mild. No constitutional effects are associated (as contrasted with weight loss, anemia, fever, etc., in rheumatoid arthritis). Involvement of the soft tissues overlying muscles consists of a painful swelling, an inch or more in diameter, which is brawny, firm and tender but does not itch or burn (in contrast with the *swelling of angioneurotic edema*). Favored sites of swellings are the bottoms of heels, finger pads, distal phalanges, flexor surfaces of forearms, thumb pads and Achilles tendons. Occasionally, a subcutaneous nodule may be palpable in the hand. Laboratory and roentgenologic findings are negative. Pathology consists of low-grade inflammation which completely subsides without residual damage. No treatment is known, although gold compounds have been used successfully.[27] The condition is chronic but may be cured spontaneously or reduced in severity in a majority over the years.

[25] Hench, P. S, and Rosenberg, E. F. Arch. Int. Med. 73:293, 1944

[26] Hench, P. S, *et al.*, (Rheumatism Review) Ann. Int. Med. 28:66, 309, 1947.

NEUROARTHROPATHY
(Neuropathic Joint; Charcot Joint)

A neuropathic joint is one associated with central or peripheral nerve lesions and characterized pathologically by extreme destruction, pronounced new bone formation and elongation of the supportive structures, and clinically by painlessness and abnormal mobility.

Etiology

Ninety per cent of cases occur in conjunction with *tabes dorsalis*, and mainly in the lower extremity. Most of the remainder are associated with *syringomyelia* and mainly in the upper extremity. Rare cases apparently are related to peripheral nerve lesions (including leprosy), various spinal cord lesions, and cerebral lesions, notably arteriosclerotic degenerative disease.

The actual mechanism of production is unknown. Eloesser showed experimentally that trauma to an anesthetic joint will result in changes comparable with a Charcot joint.[28] Unexplained, however, is involvement of a single joint in an extremity while the other equally anesthetic joints remain undisturbed.

Males are predisposed. The areas of predilection in decreasing order of frequency are: knee, foot, ankle, hip, spine, elbow, shoulder and wrist.

Pathology[29, 30]

The picture is typified by destruction, re-

[27] Boland, E. W, and Headley, N. E.: Ann. Rheum. Dis. 7:246, 1948

[28] Eloesser, L., On the nature of neuropathic affections of joints, Ann. Surg. 66:201, 1917.

[29] Luck, J. V.: Bone and Joint Diseases, p. 243, Springfield, Ill, Thomas, 1950

[30] Ghormley, R. K. (ed.): Orthopedic Surgery, p. 375, New York, Nelson, 1938.

Roentgenographic Findings

At the time of the initial swelling, the roentgenograms are nonrevealing. Gradually, over the ensuing weeks the joint surfaces become denser and yielding at points of bony contact and pressure. The surfaces disintegrate. Bone shadows appear in the periarticular soft tissues. The bone architecture beneath the articulating cortex becomes sclerosed. Free ossific bodies appear within the joint cavity. Large marginal exostoses develop. Pathologic fractures heal with considerable callus. In the knee, the medial femoral condyle is often the site of earliest changes. In the foot, the midtarsal joints are affected most frequently. Disintegration of the ankle may follow a Pott's fracture. Subluxation, dislocation and deformity are late findings.

Treatment[31]

In the acute stage, the joint should be shielded from trauma of ordinary motion and weight-bearing. The fluid should be aspirated and the limb immobilized in a cast. The limb is elevated until swelling is reduced. After the acute stage, the articulating bones are hardened and can be used with the proper support. The following measures are useful:

Knee. A straight caliper with an ischial fitting ring. A leather corset about the knee may be added. In severe cases, arthrodesis is best, but difficult to obtain. *Procedure:* A preliminary operation to improve circulation consists of *drilling holes* through the areas of sclerosis.[32] Several weeks later arthrodesis is done. The irregular sclerotic surfaces are resected, and two flat ends are apposed Two *cross grafts* are inserted from femur to tibia after the method of Brittain The *compression* apparatus of Charnley is applied, and the limb is suspended on a Thomas splint. Compression is gradually increased over the ensuing weeks until early fusion can be demonstrated. This is followed by *prolonged immobilization* in a cast which is discontinued only when good bony bridging has occurred.

Ankle. A brace or ankle corset is used for mild cases, and crutches may be worn. Arthrodesis is best. The principles of drilling, grafts, fixation, compression and prolonged immobilization should be followed.

Foot. Destruction is in the distal tarsal area where fusion is almost impossible. Special shoes with steel arch supports plus a cane will permit ambulation. The danger of trophic ulcers with secondary infection is great. Weight-bearing should be limited or avoided and a metatarsal bar worn on the shoe. Extreme destruction warrants amputation.

Hip. Extreme destruction followed by subluxation with loss of stability is usual. A subtrochanteric osteotomy of the Shanz type with 30° of abduction provides stability.

Spine. A corset or jacket may be worn. However, progression of deformity is usual. A spine fusion is indicated.

Elbow. Disability is minimal. A leather-hinged corset which permits only flexion-extension movement improves function.

Neurogenic Arthropathy Associated With Diabetes Mellitus.[33] In diabetic patients of advanced age, severe arteriosclerosis, and poorly regulated diabetes of long standing, a Charcot type of arthropathy may occur. The foot is prone to involvement, a painless swelling without inflammatory signs appearing in the tarsal area. Roentgenograms reveal destruction and fragmentation. Trauma frequently initiates the condition. Additional symptoms of night pain and paresthesias and findings of loss of reflexes and vibratory sense imply peripheral nerve changes. The spinal fluid protein and cell count are increased. Inadequate circulation makes conservative treatment mandatory.

SUPPURATIVE ARTHRITIS
(Pyogenic Arthritis)

An acute arthritis due to specific bacteria and productive of purulent exudate is designated a suppurative arthritis.

Etiology

The following are the definitely known factors:

Predisposing Causes. Children are most commonly affected. An infective focus fre-

[31] Key, J A : Treatment of Charcot's joint, Urol & Cutan. Rev 49 161, 1945

[32] Soto-Hall, R.. Fusion in Charcot joint of the knee, Ann. Surg. 108·124, 1938.

[33] Lippman, E. M., and Grow, J. L : Neurogenic arthropathy associated with diabetes mellitus, J. Bone & Joint Surg. 37A:971, 1955.



Fig. 126. Neurotropic joint.



Fig. 127. Charcot knee.

Fig. 128 Charcot ankle and foot.

Roentgenographic Findings

At the time of the initial swelling, the roentgenograms are nonrevealing. Gradually, over the ensuing weeks the joint surfaces become denser and yielding at points of bony contact and pressure. The surfaces disintegrate. Bone shadows appear in the periarticular soft tissues. The bone architecture beneath the articulating cortex becomes sclerosed. Free ossific bodies appear within the joint cavity. Large marginal exostoses develop. Pathologic fractures heal with considerable callus. In the knee, the medial femoral condyle is often the site of earliest changes. In the foot, the midtarsal joints are affected most frequently. Disintegration of the ankle may follow a Pott's fracture. Subluxation, dislocation and deformity are late findings.

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Exciting Causes. *Staphylococcus aureus* or *albus* and hemolytic streptococcus most commonly. Less commonly the pneumococcus, the meningococcus, the gonococcus, typhoid, etc.

Mode of Infection. Often hematogenous from infective focus. Less commonly, spread from an adjacent focus, especially osteomyelitis, or by direct introduction through a wound.

Areas of Predilection. Knee, hip (especially in infants), shoulder.

Pathology

Basically, the condition is an acute synovitis which varies in degree, depending on the virulence of the organism and the resistance of the tissues. In a mild case the synovium is congested, edematous, and infiltrated with polymorphonuclear leukocytes. Serum exudes into the joint cavity where it admixes with an increased amount of synovial fluid and deposits flakes of fibrin over the inner lining of the joint. The serous fluid is clear or slightly opaque and contains a slight amount of polymorphs The synovial sac is greatly distended by the large amount of fluid The condition is designated as *serous arthritis*, or, when precipitation of fibrin is excessive, as *serofibrinous arthritis*. When inflammation is more intense, congestion, edema and leukocytic infiltration are greater in degree. Areas of vascular thrombosis and focal necrosis occur. The joint exudate contains a large number of polymorphonuclear leukocytes, as high as several hundred thousand per cubic millimeter, and therefore is very opaque, thick, and gray or yellowish gray in color. Proteolytic enzymes originating from the polymorphonuclear leukocytes dissolve the articular cartilage and may even erode the bone. The intense intra-articular pressure causes necrosis and destruction of intra-articular soft tissues and capsule, the exudate erupting into the surrounding soft tissues and even through the skin. This is the typical *purulent arthritis* characteristic of staphylococcus infections. When hemolytic streptococcus is the causative organism, the infection is fulminating and quickly destructive. The synovial inflammatory signs are intense, vascular thrombosis and necrosis are extreme, and hemorrhagic extravasations are seen. The defenses are not as adequate as in the case of staphylococcus so that polymorphs are not as numerous. The exudation into the joint consists of a bloody serous fluid typical of *serosanguineous arthritis*. The danger of a fatal septicemia is greatest in this type.

Healing in the mild serous form takes place by resolution. The serous exudate is resorbed, and inflammation of the synovium subsides, the joint returning to normal. The more serious destructive arthritis requires not only the resorption of purulent exudate but also repair by granulation tissue (resolution and organization) which bridges the joint and eventuates in fibrous ankylosis. The capsule becomes fibrotic, thickened and inelastic. Loss of articular cartilage exposes the bone to mechanical trauma resulting in degenerative arthritis.

Acute suppurative arthritis of the hip which occurs most often in infants presents an unusual situation. The femoral head is composed almost entirely of cartilage which lies completely within the articular cavity. The distention of the capsule by the exudate shuts off the circulation to the head and the enzymatic action dissolves the cartilage. In consequence, the femoral head disintegrates, and subluxation and dislocation result. Because the ossification center does not normally appear until the 6th or 7th month and may be delayed by the infection, recognition of a pathologic dislocation of the hip may not be recognized until the walking age.

Clinical Picture

A history of antecedent trauma and infection may be obtained.

Symptoms. Pain gradually increases in intensity over several hours, eventually becoming excruciating. It is accentuated by joint movement and, in the case of the lower extremity, by weight-bearing. Constitutional symptoms of an acute infection include chills, fever, sweats, malaise, anorexia and, in infants, nausea and vomiting.

Findings. The patient limps if the lower extremity is involved. Usually, only one joint is affected. The joint is swollen, red, warm and tender throughout, and the position is one of partial flexion. The swelling consists of increased joint fluid which may obliterate the

markings. In the case of the knee, the patella is floating. In a gonorrheal arthritis, a peculiar soft tissue edema surrounds the joint. The muscles are in protective spasm. The temperature is elevated, spiking daily as high as 104°.

In the hip the thigh is held in flexion, abduction and external rotation, for this is the position of greatest relaxation of the capsule. Pain is referred along the inner side of the thigh to the medial aspect of the knee.

Laboratory Findings

Aspirated Joint Fluid. This is serous, serosanguineous, or frankly purulent. Microscopically, the offending organism may be identified. The fluid is cultured, and the bacteria are tested for susceptibility to antibiotics.

Blood Count. A high leukocytosis, polymorphs predominting

Increased Sedimentation Rate

Positive C-Reactive Protein

Roentgenographic Findings. Films are generally negative at the outset. However, ballooning of the synovial sac may be interpreted from the rounded soft-tissue outlines and density peculiar to purulent exudate. If the infection persists, osteoporosis of all bones adjacent to the joint takes place. With destruction of cartilage, narrowing of the joint interval occurs. Degenerative arthritis supervenes as a late sequel. A neighboring osteomyelitis may be revealed.

The *obturator sign* is diagnostic of hip involvement. A soft-tissue shadow is normally seen over the lateral wall of the pelvis and medial to the acetabulum. When the joint is distended by fluid, the shadow bulges inwardly, becoming very prominent by comparison with the opposite side. The obturator internus is presumably the structure involved. The sign affords a valuable diagnostic aid in infants.

Treatment

Drainage. In suppurative joint disease immediate and free, dependent drainage is mandatory. The proteolytic action and pressure of the exudate is very destructive. Fatal septicemia, bone infection and, if infection is overcome, a disabling ankylosis are complications which can be avoided by early diagnosis and instituting drainage. Aspirations are mentioned only to be condemned. The exudate rapidly reaccumulates, and destruction continues. Repeated insertion of a needle may introduce additional infection. One should not judge an infection as "mild" and depend on aspirations and antibiotics. Although many such cases can be handled by such conservative treatment, an occasional case will progress and threaten local structures and even life itself. Energetic surgical measures should not be executed as an afterthought. Like any other surgical infection, given the opportunity the body defenses will eliminate the infection. The joint is opened on two sides to provide through-and-through gravity drainage. Gutta-percha drains may be inserted to prevent premature closure and are removed in 24 to 48 hours. The synovium is not sutured. It will close spontaneously and rapidly when infection is controlled. The only exception to surgical drainage is gonococcus infection, which responds promptly to penicillin.

Antibiotics are given in adequate amounts. Penicillin, which combats most gram-positive organisms, may be combined with streptomycin, which is effective against gram-negative organisms. One million units of penicillin and 2 Gm. of streptomycin are given in divided doses intramuscularly each day. In the meantime, several days are consumed in determining antibiotic sensitivity. The so-called broad-spectrum antibiotics as chlortetracycline, oxytetracycline, etc., may be substituted when indicated. Because antibiotics, especially streptomycin, do not readily pass through the inflamed synovium into the joint, they must be introduced directly. Through a catheter, a constant drip of a solution of the antibiotic in normal saline is maintained.

Immobilization. The limb is immobilized in a removable splint as a folded blanket and is elevated. The position of function must be maintained in the event that ankylosis ensues. Movement during the acute stage is forbidden. However, in the subacute subsiding stage the joint should be moved gently and passively each day as a precaution against formation of ankylosing adhesions. When the hip or the knee is involved, traction is advisable. It immobilizes the joint, distracts the articular surfaces, thereby reducing damage due to pressure, and affords convenience for changes of dressings.

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Supportive therapy includes fluids, transfusions, a highly nourishing diet and sedation.

The temperature usually falls to normal, drainage subsides, and the wounds close spontaneously. In rare instances, if the infection progresses unabated and constitutes a serious menace to life, amputation is indicated.

A nonfunctional ankylosis resulting from suppurative arthritis should be treated with respect. Results of arthroplasty, except in cases where the joint has been preserved, are disappointing. Arthrodesis in the position of function is best.

Pathologic dislocation of the hip can be handled by subtrochanteric osteotomy or arthrodesis.

GONORRHEAL ARTHRITIS

Gonorrhea and its complicating arthritis has become an uncommon disease since the introduction of antibiotics. However, one must be on guard to recognize and treat immediately the occasional case of gonorrheal infection because of its destructiveness to the joint. It is possible for gonorrhea to lie dormant for many years and suddenly make its appearance as a genito-urinary infection, arthritis, conjunctivitis, etc. Pelvic surgical procedures and urethral instrumentation have been known to provoke the infection. The appearance of an arthritis following these procedures should arouse suspicion.

Pathology

The process is similar to other forms of suppurative arthritis. The synovium is congested, edematous, and infiltrated with polymorphonuclear leukocytes. The articular cartilage is digested by the tryptic substances in the pus. A large amount of synovial fluid contains a preponderance of polymorphs. Sugar content of the fluid is decreased. Healing takes place by fibrosis of the synovium, and fibrous adhesions bridge the joint, resulting in ankylosis. Bony ankylosis is uncommon.

Clinical Picture

Usually within 3 weeks after an acute gonorrheal urethritis a polyarticular arthritis develops, most commonly in the knees, the wrists and the ankles. The joints become hot, swollen, reddened, tender, and distended with a large amount of fluid. The periarticular tissues present a peculiar diffuse edematous boggy swelling. Pain becomes severe. Tenosynovitis, particularly about the ankles and the wrists, is a frequent accompaniment. The temperature rises as high as 104°. A high polymorphonuclear leukocytosis and an increased sedimentation rate are laboratory findings. The complement-fixation test is not of value unless a negative test becomes positive during the course of the disease. After a few days, the arthritic symptoms and findings subside but may persist in one joint as a resistant monoarthritis. If untreated, the destruction of articular tissues eventuates in ankylosis. Typical gonorrheal arthritis is self-limited and runs its course in about 3 months.

Diagnosis

Isolation of gonococci by culture of aspirated joint fluid constitutes the most reliable diagnostic measure.

Treatment

The gonococcus is highly susceptible to penicillin. It is unnecessary to inject the antibiotic intra-articularly. The affected joints are put at rest by splinting. Daily the splints are removed for passive motion to break up soft newly formed adhesions. The position of function is sought in the event that ankylosis takes place. After the acute stage, physical therapy is instituted to restore full motion to the joint and strength to the atrophied muscles.

Ankylosed joints, particularly when destruction is not too far advanced, are suitable candidates for arthroplasty. When loss of joint surfaces has resulted in a painful degenerative arthritis or a deformity, arthrodesis in the functional position is best.

REITER'S SYNDROME
(Infectious Uro-arthritis)

Reiter's syndrome is a self-limited condition of unknown etiology consisting of a characteristic triad of urethritis, conjunctivitis and arthritis.[34, 35]

Etiology

A filtrable pleomorphic ultramicroscopic

[34] Hollander, J. L.: Medical Clinics of North America, vol. 30, p 716, Philadelphia, Saunders, 1946

[35] Paronen, I.: Reiter's disease, Acta med. Scandinav. (supp. 212) 131 1-114, 1948.

pleuropneumonialike organism, the "L" organism, has been cultured from the urethral discharge. These organisms are present in the genito-urinary tract of both male and female so that transmission by sexual contact is probable. The condition affects males between the ages of 20 and 40 most commonly. Other etiologic factors blamed but not proved include dysentery, spirochetes, atypical gonorrhea and allergy.

Pathology

The synovial tissue displays only edema and congestion with perivascular collections of round cells. Suppurative inflammation of the urinary tract involves the urethra, the prostate, the bladder, and the pelvis of the kidney. Contrary to gonorrhea, the epididymis is never affected. The conjunctiva is congested, but other eye structures are uninvolved.

Clinical Picture

This varies in severity and duration. The initial symptoms are those of urethritis, including frequency, dysuria and purulent discharge, and lasting anywhere from a few days to several months. Recurrences are common after apparent subsidence. A catarrhal conjunctivitis causing lacrimation, photophobia and discharge makes its appearance and lasts a few days to several weeks, subsiding without permanent damage. Within 2 weeks after onset of urethral discharge, an acute polyarthritis appears usually accompanied by a temperature elevation up to 101° daily. The weight-bearing joints, particularly the ankles and the knees, are most commonly involved. The joints are hot, swollen and tender. The periarticular edema characteristic of gonorrheal arthritis is not found Synovial fluid is abundant and clear. Keratotic and pustular skin lesions are found in the more acutely ill patients Mucous membrane ulcerations, especially of the oral cavity and the glans penis, are frequent.

The condition is self-limited. The acute stage lasts 4 to 6 weeks, and the entire course about 6 months

Laboratory Findings

1. **Leukocytosis up to 12,000**
2 **Increased sedimentation rate,** reaching its peak at 6 weeks
3. **Purulent urethral exudate** in which gonorrheal organisms are absent. Culture may reveal "L" organisms.
4. **Synovial fluid cell count 9,000 to 53,000,** lower than in synovial pyogenic infections
5. **Gonococcus complement-fixation tests are negative.**
6. **Agglutination reaction** of serum for pleuropneumonia organisms may be present in significant titres.

Roentgenologic Findings

Moderate osteoporosis about the involved joints appears during the acute stage of arthritis.

Treatment

Streptomycin and oxytetracycline are effective for joint infection. Hot packs are applied to the joints. Symptomatic treatment is prescribed for urethritis and conjunctivitis. Prostatic massage tends to cause exacerbation of symptoms. ACTH and cortisone are very effective in eliminating symptoms, but recurrence is the rule after the hormone is stopped. Without treatment the condition is self-limited, all symptoms subsiding without permanent residuals.

HEMOPHILIC ARTHRITIS

Hemophilia is a hereditary condition characterized by the occurrence of hemorrhages, most commonly in a joint, as a result of prolonged coagulation time of the blood.[36-39]

Etiology

It is transmitted as a sex-linked mendelian recessive from the male through the unaffected female offspring to another male. A *prolonged coagulation time* is the cause of the hemorrhagic diathesis. Experimental evidence suggests a deficiency of a substance which normally is present as a Phase I precursor of thromboplastin. This factor can be supplied by whole blood, fresh plasma, or plasma free of platelets. Serum lacks this substance. The factor can be precipitated from plasma by acid

[36] Hollander, J L , *et al.* (ed): Comroe's Arthritis, Philadelphia, Lea & Febiger, 1953

[37] Ghormley, R K., and Clegg, R S : Bone and joint changes in hemophilia, J. Bone & Joint Surg 30A·589, 1948

[38] Key, J A Hemophilic arthritis, Ann Surg. 95· 198, 1932

[39] Winston, M E Hemophiliac arthropathy of the hip, J. Bone & Joint Surg 34B:412, 1952

and is available as Cohn's plasma fraction I. The bleeding is initiated by trivial trauma or may appear to be spontaneous. Other blood tests as bleeding time, platelet count and prothrombin time are normal.

Clinical Picture

A history of bleeding as a result of trivial trauma is typical. The patient is a male who describes many incidents of uncontrollable hemorrhage following tooth extraction, minor cuts and bruises, and most commonly into and about joints. Acute hemarthrosis occurs in one joint as a rapid effusion with marked swelling developing within a few minutes to several hours. The knee is involved most commonly, less often the ankle, the elbow and the hip. The swelling follows the outline of the synovial cavity. Pain is severe, the joint feels warm, and its position is one of flexion. This acute phase lasts for a few days to several weeks. After the blood is absorbed, the chronic synovitis persists for many weeks and months as a tender, sore, painful and swollen joint. Recurrences are common. With each attack the joint becomes progressively limited in motion, thickened and deformed, usually in a position of flexion. Degenerative arthritic symptoms supervene, but ankylosis which is fibrous, occasionally bony, eventually results in fixed flexion deformity The musculature about the joint becomes atrophied, the final picture of deformity resembling that of rheumatoid arthritis.

Large bulbous hemorrhages may develop along the shafts of long bones, producing characteristic hemophilic pseudotumors. These large accumulations of blood may destroy the overlying muscles and skin, with terminal external hemorrhage; they may erode the bony cortex, destruction of the shaft by intramedullary hemorrhage or by erosion from a *subperiosteal hemorrhage may occur It is im*possible to state the origin of a pseudotumor, whether by extension from an adjacent joint, from subperiosteal vessels, or from intramedullary hemorrhage.

When hemarthrosis occurs in the hip before puberty, changes occur in the epiphysis similar to Legg-Perthes' disease, resulting in stiff-hip limp and restriction of abduction and internal rotation. Bleeding into the hip in adulthood results in changes comparable with malum coxae senilis.

Roentgenologic Findings

The initial acute hemarthrosis displays a bulged-out capsule and increased density due to blood. Opacities in the soft tissue signify the presence of blood extra-articularly. In chronic cases after several recurrences the joint interval narrows as a result of thinning and destruction of articular cartilage. The articular cortex becomes interrupted and the subchondral trabeculae replaced by cysts and cavities supposedly due to intramedullary hemorrhages. The articulating bones are characteristically demineralized. Eventually, the mechanical imperfection of the joint leads to hypertrophic degenerative changes and osteophytes. At the site of a pseudotumor, a bulbous opacity lies against a bony defect which varies from a slight saucer-shaped erosion to deep cysts or cavitations which destroy the shaft.

In children, the epiphyses about the joint may be enlarged and misshapen. At the hip, increased density, flattening, and fragmentation suggestive of Perthes' disease are seen.

Pathology

Blood mixed with synovial fluid acts as an irritant to the synovial membrane. This has been proved experimentally in animals. The synovial membrane becomes inflamed and undergoes fibrosis and villous formation. Removal of the blood is through the synovium where in the subsynovial stratum can be seen disintegration of erythrocytes and macrophages filled with blood pigment. Reactive granulation tissue forms and organizes into scar tissue. The granulation tissue forms a pannus which extends over and absorbs the articular cartilage at the margins. Probably as a result of lack of synovial fluid nutrition, the articular cartilage throughout the joint becomes soft, yellowish and degenerate. It deteriorates or becomes eroded by pressure in *an irregular fashion to produce a characteris*tic maplike appearance The articular cortex becomes thinned and worn through. Subchondral hemorrhages in the porotic bone produce cysts by engulfing the fine trabeculae which, deprived of circulation, necrose and absorb. It is possible that blood may have some chemical absorptive effect on bone The irregularity of the joint leads to secondary degenerative arthritis. Adhesions eventually bridge the joint, resulting in a fibrous ankylosis which may ossify.

Gradual onset over hours of *swelling, pain* and *limitation of motion* implies increased effusion with a minimum of blood

Acute onset over minutes to an hour indicates intra-articular hemorrhage.

Objective findings: *Swelling* is mainly intra-articular, due to fluid accumulation. In the knee, the patella is ballotable. *Edema* and *tenderness* are localized over the site of trauma. The position is one of *semiflexion*. *Motion is limited* in all directions, due to obstruction of fluid. Abnormal mobility in a lateral direction indicates torn collateral ligaments. Evidence of other soft-tissue injury should be sought (These findings are described in chapters devoted to specific regions.)

Laboratory Findings

Aspiration of joint fluid which varies from a clear amber to bloody. Its removal, in the absence of complicating soft-tissue injuries, should restore complete motion to the joint.

Roentgenograms. Early films are negative. When the fluid is frankly bloody, a characteristic soft tissue density may be seen. Air arthrograms are used to identify a piece of cartilage which is torn and displaced from the articular surface or a meniscus The changes due to developing osteoarthritis gradually appear over many months and take place at a faster rate than in other joints.

Treatment

The joint is placed at *rest* by a splint of metal or plaster. A *compression dressing* and an *icebag* retard hemorrhage. *Muscle-setting exercises* are practiced, while the splint is worn, to reduce the muscle atrophy and to prevent periarticular adhesions Removal of fluid by aspiration to relieve pain or the introduction of compound F is not justified. Infection is a definite danger. Bloody joint fluid is an excellent medium for the growth of organisms introduced by the aspirating needle. As the swelling is subsiding, *gentle passive exercises* and later *active non-weight-bearing exercises* are practiced. Mobility of the joint and integrity of the musculature are maintained. A severely injured joint should not be subjected to weight-bearing until all signs of inflammation have subsided. This precaution against damage to articular surfaces may require several weeks. *When recurrent effusions occur with repeated attempts at weight-bearing, an intra-articular mechanical impediment should be suspected.* Arthrography may demonstrate a cartilaginous joint mouse. Occasionally, exploratory arthrotomy may be necessary to identify the cause. One should not wait until degenerative changes have set in before resorting to such measures. When a periarthritis has resulted after an acute traumatic arthritis, physical therapeutic measures are required. Forceful stretching should be avoided, as tearing of soft tissue and formation of more adhesions further restrict motion.

When ambulation with full weight-bearing is resumed, muscle weakness causes joint instability which may lead to strains and reactive synovitis An elastic compression bandage should be worn.

PSORIATIC ARTHRITIS

Occasional cases of psoriasis develop arthritis which is clinically similar but distinct from rheumatoid arthritis.[40, 41]

Clinical Picture

Skin changes usually precede the appearance of joint symptoms by months to several years, but rarely skin and joints may be involved simultaneously. Disease of the nails similarly antedates the arthritis. The initial symptoms usually appear in a single joint, developing insidiously or suddenly, and the distal interphalangeal joint of a finger or a toe is often the site of earliest involvement. A finger or a toe can swell up within a few hours, become tense, livid and shiny and simulate the clinical appearance of gout. The syndrome evolves in attacks and remissions. During exacerbations, severe transient migratory aches and pains develop in several joints, particularly the spine. In the beginning, the involved joint or joints return to normal between attacks, but later some residual damage with some deformity and limitation of motion results.

In the hand, the distal interphalangeal joints and later the proximal interphalangeal joints, but practically never the metacarpophalangeal joints, are involved. Less often the

[40] Sherman, M : Psoriatic arthritis, J. Bone & Joint Surg. 34A:831, 1952.

[41] Bauer, W., Bennett, G. A , and Zeller, J. W.: The pathology of joint lesions in patients with psoriasis and [illegible]

wrist is involved, usually the ulnar aspect, and often associated with a tenosynovitis. Never does the typical "rheumatoid" ulnar deviation deformity develop.

In the foot, the distal and then the proximal interphalangeal joints and finally the metatarsophalangeal joints are affected. The toes become displaced dorsally and proximally. The tarsus and the ankle are spared.

Characteristically, the disease progresses joint by joint until many joints, including the knees, are involved.

Roentgenographic Findings

The bone is never atrophic. Early, marginal erosions are observed at the interphalangeal joints. Later, joint destruction is apparent and extends along the surfaces of the phalanges as scalloped indentations. The ends of the phalanges appear "gnawed away" and thinned. An overgrowth of bone at the tendon insertions produces a "cuplike" deformity. The changes show a predilection for the distal joints of the fingers and the toes. Rarely, and much later than is noted in rheumatoid arthritis, bony ankylosis occurs.

Pathology

Basically, the process consists of chronic inflammation with edema and round cells followed by replacement fibrosis. The inflammatory tissue erodes the cortex and the cartilage at the periphery of the articular end of the phalanx. The destruction extends centrally and exposes the cancellous bone to the joint. Dense fibrotic tissue eventually occupies the entire joint. The inflammatory tissue extends along the surface of the shaft which is eroded from without and causes the characteristic scalloped appearance. Contrary to rheumatoid arthritis, one never observes pannus formation, dense accumulations of round cells, and osteoporosis. Rarely, complete bony ankylosis is seen, but dislocation and subluxation are common.

Differentiation From Rheumatoid Arthritis

The only similarity is an elevated sedimentation rate. Otherwise, psoriatic arthritis does not exhibit fever, leukocytosis, lymphadenopathy, ulnar deformity of the hand, intrinsic muscle contracture, skin atrophy, iritis, cardiac changes, or subcutaneous nodules.

Treatment

A good clinical remission can be effected by relatively large doses of cortisone (200 to 300 mg. daily) or ACTH (100 to 200 mg. daily). Then the dose is reduced to the amount necessary for maintenance. After 6 to 10 weeks, the drug is withdrawn, the remission lasting from 6 weeks to 6 months.

ALTERATIONS IN THE COMPOSITION AND THE FUNCTION OF SYNOVIAL TISSUES RELATED TO TUMOR FORMATION[42]

The intimal layer of the articular capsule is made up of specialized mesenchymal cells that have the capacity of regulating the passage of substances between the articular and the vascular fluids and of secreting mucin. Similar tissues line the tendon sheaths and the bursal spaces. Under certain mechanical influences, e.g., irritation over a bony prominence, a synovial lined bursal space will form. The synovial lining of a joint varies from dense connective tissue lined by inconspicuous flattened cells to a membrane comprised of many layers of round, oval, or cubical cells which are supported by a loosely textured and highly vascular connective tissue. These latter areas may contain recesses or crypts, as well as permanently formed papillary projections called villi. With flexion and extension of the joint, temporary folds are also produced in the synovial-lining layer of cells and its supporting subsynovial tissues. Another function is the transportation of colloidal and particulate matter across the synovial membrane. Continuous or intermittent bleeding into the articular cavities leads to extensive storage of hemosiderin in the phagocytes of the subsynovial tissues or in the synovial-lining cells. Lipoids and other phagocytosed substances may also accumulate in these tissues under certain abnormal circumstances.

Under the influence of injury and persistent irritation, neoplasmlike overgrowths of the synovial tissues will result. Some lesions are heavily pigmented with hemosiderin, while

[42] Bennett, G A: Malignant neoplasms originating in synovial tissues, J. Bone & Joint Surg. 29:259, 1947.



FIG. 129. Xanthoma cells or foam cells. They are large, polyhedral, contain a small, central, usually pyknotic nucleus and transparent, foamy cytoplasm. Lipoid bodies are dissolved out in H-E stains and are demonstrable by fat stains. Foam cells exist in varying proportion with fibroblasts and collagenous fibrils. When fibrous tissue is abundant, xanthoma cells are enveloped and difficult to demonstrate, the lesion being termed a *fibroxanthoma*. The following are characteristics of a fibroxanthoma: (1) *foam cells*; (2) *fibrocellular stroma* with short, blunt, spindle-shaped cells containing a pale, slightly elongated nucleus; (3) *multinucleated* giant cells of the epulis type containing 5 to 40 nuclei; (4) *blood pigment,* coarse yellow or brown pigment of hemosiderin scattered throughout the groundwork, situated principally intracellularly in macrophages or foam cells; (5) *blood vessels,* mainly small abundant capillaries; and when xanthomatous tumors develop within the synovium of joints (nodular synovitis), there is added (6) *synovial villous formation.* (DeSanto, D. A., and Wilson, P. D.: Xanthomatous tumors of joints, J. Bone & Joint Surg. 37:531)

others may contain large amounts of lipoid alone or in combination with hemosiderin. The structural peculiarities of synovium are reproduced in the new growth. Therefore, fibrous connective tissue elements are the main component in one tumor, whereas another would display prominent tufting or villous overgrowths. In still another, the lining cells are prominent and multiply so that several layers of cells are seen. These new growths can occur also in regions far removed from joints, as the mesenchymal tissue always has a synovial forming capacity.



FIG. 130. Pigmented villondular synovitis. Note nodular density in the suprapatellar pouch demonstrated by air arthrogram.

PIGMENTED VILLONODULAR SYNOVITIS

(Synovial Xanthoma; Villous Synovitis)

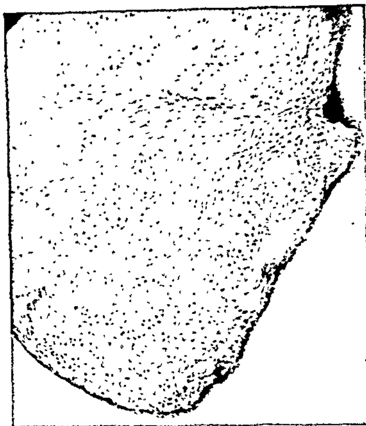
Pigmented villonodular synovitis is the name applied by Jaffe and his associates to an idiopathic villous overgrowth and pigmentation of the synovial membrane of a single joint. Many names have been applied to these xanthomatous lesions which are characterized by their yellow or yellowish-brown color due to deposits of cholesterol and hemosiderin *Since bursae and tendon sheaths are related* to synovium in their origin, they too are the site of xanthomatous growths.[43]

CLINICAL PICTURE

Age. Usually the young adult from 20 to 40. The *diffuse villous form* occurs in the 2nd and the 3rd decades; the *circumscribed nodular growth* occurs in the 3rd to the 5th decades

[43] Jaffe, H L, Lichtenstein, L, and Sutro, C J. Pigmented villonodular synovitis, Arch. Path. 31:731, 1941.

PLATE 12. Chondromatosis. Note the synovial lining about the cartilage. (× 85)



Sex. Males most commonly.

Area of Predilection. Knee joint, a few in the ankle. Rare elsewhere.

Trauma. History occasionally obtained, relationship doubtful.

Duration of Symptoms. From 1 day to many years.

Course. Usually gradual onset of pain, mild to moderate, intermittent and associated with a limp. Mechanical interference causes stiffness, locking, limitation of motion usually in extension, and a snapping sensation at times. In the more localized form, the symptoms are mild and develop very gradually; therefore, as a rule, patients do not seek advice early.

Findings. A soft-tissue swelling enlarges the entire joint. The effusion may be pronounced, the patella floating. Moderate generalized tenderness. *In the localized form,* a palpable small tumor may suggest a joint mouse. Usually, in this type the tumor is not palpable, and only after air arthrography or on exploration is the true nature of the disease defined. The swelling is slight and intermittent. Effusion is minimal.

Aspiration. A thick orange-brown fluid containing cholesterol in large amounts is pathognomonic. In the localized form, however, the effusion is not abundant, is straw colored and sterile.

Laboratory Findings. These are not diagnostic. The blood cholesterol is at the upper limits of normal. No alteration of the cholesterol-cholesterol ester ratio exists.

Roentgenologic Findings

Direct films of the joint are usually negative or may display general soft-tissue enlargement. Occasionally, moderate decalcification from tumor pressure is noted. An air arthrogram reveals, in the diffuse type, a bubbly flocculent effect within the synovial cavity. In the localized type, a soft-tissue shadow encroaches upon the synovial pouches, most commonly in the posterior part of the suprapatellar area.

Pathology

The joint may be distended by a chocolate-colored mass with small or large, soft or hard, nodular masses on a base of fibrous connective tissue having the consistency of rubber. The cut surface resembles fine sponge rubber and displays a mixture of gray and pale yellow color. Microscopically, fibrous connective tissue contains many cells, the nuclei of which are vesicular but vary in size and shape. Some are oval, some spindle-shaped, and others polyhedral. Scattered yellow pigment may be contained in large oval or polyhedral cells, resembling large monocytes or histiocytes. Cells containing lipoid material, the

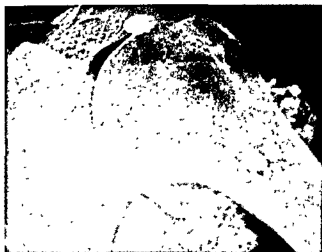




Fig. 131. Osteochondromatosis. (*Left*) Shoulder joint (*Right*) Ankle.

foam cells, occur in varying amounts. Occasional giant cells of the epulis type occur about areas of destruction and debris. The synovium is thrown into folds, the stroma is edematous and markedly vascular, perivascular cuffs of lymphocytes may be seen, and in the later stages when fibrous tissue proliferation occurs, the cells are squeezed out, becoming sparse, and the vascularity is lessened.

Theories of Pathogenesis[44]

Jaffe, Lichtenstein and Sutro favor the theory that xanthoma represents an inflammatory response, the causative agent being unknown. The xanthoma or foam cells are derived from the macrophages of the reticuloendothelial system and are modified to contain cholesterol They do not believe in bacterial, lipoid meta-

[44] Minear, W. L Xanthomatous joint tumors, J. Bone & Joint Surg 33A 451, 1951

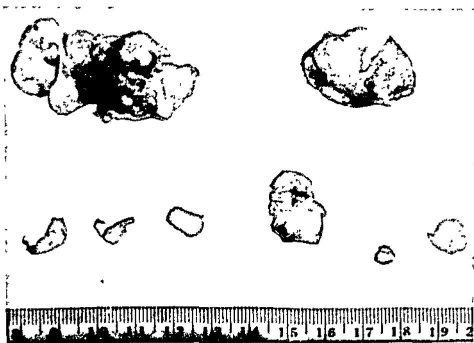


Fig. 132. Osteochondromatosis. These loose bodies were recovered from a knee joint.

bolic and hemorrhagic causes, because the condition cannot be reproduced experimentally. They reject the theory of Geschickter and Copeland who state that the lesion develops through proliferation of osteoclasts in associated sesamoid bones. However, recently Young and Hudacek have experimentally produced the growth in dogs.[45]

Treatment

Surgical excision is indicated. Recurrences are common but are adequately handled by x-ray irradiation. No malignant degeneration has ever been reported.

OSTEOCHONDROMATOSIS

In this disease, cartilaginous or osteocartilaginous bodies develop in the synovial membrane of joints or their communicating bursae.

Etiology

The condition is considered to arise from embryonic rests. Both synovial membrane and articular cartilage develop from the same mesenchymal tissue.

Pathology

Hyaline cartilage forms in the stratum synoviale of the synovial membrane, particularly at the points of reflection of the membrane. Multiple isolated areas of synovium are affected so that a tremendous number, even hundreds, of spheroid cartilaginous nodules protrude into the joint cavity. Each cartilaginous body may remain unchanged; or it may become calcified; or the calcified cartilage may be transformed to bone, particularly at its center, by metaplasia or by the process of endochondral ossification. The body gradually is extruded into the joint cavity where it is attached, at first, by a synovial pedicle. Nutrition carried through the pedicle enables both cartilage and bone to hypertrophy. The body may be torn free from its attachment, its bony center undergoing aseptic necrosis. However, it continues to grow in size by hypertrophy of its cartilaginous covering which derives its nutrition from synovial fluid. The repeated trauma to the articular surfaces by these loose bodies causes multiple erosions and eventually degenerative arthritic changes.

[45] Young, J. M., and Hudacek, A. G.: Experimental production of pigmented villonodular synovitis in dogs, Am. J. Path. 30:799, 1954.

Clinical Picture

Sex. Males predominate

Age. 30 to 50

Location. In order of frequency: knee, elbow, ankle, hip and shoulder

Symptoms. Dull ache, stiffness, transient locking episodes, grating sensations and, in the case of the knee, a giving-way

Findings. Generalized joint tenderness, thickening of the soft tissues through which the nodules may be palpable, and marked audible and palpable crepitus

Roentgenographic Findings

Only when the bodies are calcified or ossified are they visible on a direct film. Even so, because many are composed only of cartilage, the number is always much greater than one would suspect from the film. When all bodies are chondromatous, air arthrography is necessary for visualization.

Treatment

Loose joint mice must be removed immediately to halt further damage to articular surfaces. The patient should be forewarned that a certain amount of degenerative arthritis is already present and may cause residual symptoms. In addition to removal of loose bodies, a complete synovectomy is performed. All communicating bursae, as demonstrated by air arthrography, are also excised. As a general rule, the menisci are damaged, requiring removal. The outlook for permanent cure is excellent.

SYNOVIOMA

(Synovial Sarcoma; Synovial Sarcomesothelioma; Cancerous Synovial Tumor)

The normal synovial membrane consists of an *intima* which varies from a multilayered membrane of round, oval or cuboidal cells to an incomplete lining of flattened cells; and a *subintima*, a supporting layer of connective tissue which varies from loose, vascular tissue to dense fibrous connective tissue. When a multilayered type of membrane rests upon a



PLATE 13. Synovioma (× 185). Features include slitlike spaces lined with flattened inconspicuous cells, glandlike spaces lined with larger, multilayered cells, homogenous pink-staining substance in spaces, tendency toward tufting, and sarcomatous appearance of fibroblasts (irregular size, shape, staining, distribution, dense-staining nuclei and frequent mitoses).

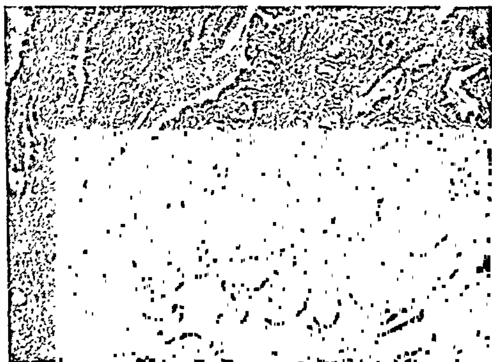


PLATE 14. (Synovioma (× 120). Pseudoadenomatous appearance. The lining cells are cuboidal and columnar. In any tumor, features of glandlike spaces, tufting and fibrosarcomatous changes should be sought throughout the tumor. One or more features may predominate.

loose supporting stroma, tufts or villi tend to form. A malignant tumor may arise from any one or all of these elements. The supporting layer may be the origin of a tumor indistinguishable from fibrosarcoma. A tumor arising from the intima displays flattened synovial or adenomatouslike elements mixed with fibrosarcomatous features. In any one tumor, either the adenomatous or the fibrosarcomatous element may predominate, but as a rule the tumor is composed of a mixture of both. The synovioma is a slowly growing malignant tumor occurring in juxtaposition to and often attached to synovial tissue but almost invariably lies outside the joint.[46-49]

[46] Bennett, G. A.: Malignant neoplasms originating in synovial tissues, J. Bone & Joint Surg. 29:259, 1947.

[47] Berger, L.: Synovial sarcoma in serous bursae and tendon sheaths, Am. J. Cancer 34:501, 1938

[48] Haagensen, C. D, and Stout, A. P.: Synovial sarcoma, Ann. Surg. 120.826, 1944.

[49] Tillotson, J. F, McDonald, J. R, and Janes, J. M.: Synovial sarcomata, J. Bone & Joint Surg. 33A:459, 1951.



Fig. 133. Synovial sarcoma removed from the popliteal space. The tumor was indurated and, although well localized, was densely adherent to the surrounding structures.

Clinical Picture

Age. Young adults, rare after 40

Area of Predilection. Lower extremity, especially about the knee

Position. In soft tissue outside of joint

Symptoms and Findings. Painful swelling begins in a periarticular or peritendinous site. It increases slowly over years before the swelling is large and pain severe. Swelling is firm or soft, moderately tender.

Course. Very slow, metastasizes eventually to the lungs.

Roentgenographic Findings

Soft-tissue technic reveals a rounded, lobulated shadow. Occasionally, stippling is observed if tumor contains small areas of calcification.

Pathology

Gross Pathology. Grossly, the tumor can rarely be identified as a synovioma, since a synovial attachment is difficult to find. The tumor is sharply circumscribed, rounded and lobulated. As it grows expansively, it compresses the surrounding tissue which become attenuated to form a pseudocapsule. Often the tumor is densely adherent to adjacent structures. Occasionally, it may be possible to trace a connection with a tendon sheath or joint. The tumor seldom forms within a joint cavity. Instead, it lies in close proximity to tendons and tendon sheaths, bursae and joint capsules. The cut section of tumor is firm, grayish pink, and often contains dark blood-stained or yellowish-brown areas of necrosis.

Microscopic Pathology. Three basic patterns indicating a synovial origin are seen: (1) *Formation of tissue spaces.* These vary from slitlike clefts lined with flattened cells to well-formed glandular spaces lined with cuboidal or columnar cells, often multilayered. A homogenous mucinouslike fluid often occupies the slitlike clefts. (2) *Formation of cell tufts.* Compact groups of oval or polygonal cells are arranged in tuftlike formations, occurring either in the solid portions of the tumor, or as papillary projections extending into the glandlike spaces. (3) *Epithelial-like cells* on a supporting stroma of compact tissue. The cells are elongated and contain small dark nuclei.

Evidence of malignity is observed in the fibrosarcomatous stroma. Occasionally, the tumor consists almost exclusively of fibrosarcoma and only after an exhaustive search for synovial elements is the true origin of the tumor revealed. Basically, the tumor is identified by: clefting, tufting, villous formations and glandlike spaces with secretion.

Prognosis and Treatment

The tumor is very slow-growing and metastasizes late. Local excision is inadequate. Radical amputation is indicated.

12

Orthopaedic Neurology

ANATOMY OF THE SPINAL CORD[1]

The spinal cord, a direct downward continuation of the medulla oblongata, starts at the upper border of the atlas and ends at the lower border of the first lumbar vertebra as the conus medullaris. Though cylindrical, it is slightly flattened in its anteroposterior diameter. Corresponding to the large nerves supplying the upper and the lower limbs, we find a cervical enlargement from Cervical 3 to Thoracic 2, and a lumbar enlargement from Thoracic 9 to 12. From the lowermost end of the spinal cord—the conus medullaris—extends a delicate median prolongation, the filum terminale interna, which ends with the dural sac at the second sacral vertebra. Its extradural prolongation—filum terminale externa—ends at the coccyx.

The spinal cord is enveloped by the dura, the arachnoid and the pia mater. External to the dura is the epidural space filled by a thin layer of fat, areolar tissue and veins. The arachnoid and the subarachnoid spaces are filled with fluid which cushions the spinal cord. The pia mater intimately surrounding the spinal cord also has lateral extensions to the inner dural surface These are equally spaced between nerve roots and are known as dentate ligaments.

In fetal life the spinal cord fills the entire length of the vertebral canal. and the spinal nerves run in a horizontal direction. As the vertebral column elongates with growth, the spinal cord is drawn upward. and therefore the roots assume an increasingly oblique and downward direction toward their foramina of exit, forming at its lowest portion the cauda equina.

The spinal nerves emerge from the spinal cord in pairs: 8 in the cervical region, 12 in the thoracic region, 5 in the lumbar region, 5 in the sacral region, and 1 pair of coccygeal nerves, making a total of 31 pairs of spinal nerves. These also correspond to varying segments of neuromeres of the spinal cord.

[1] From Ciba Clinical Symposia, August-September, 1949.

Plexuses. Each spinal nerve has an anterior and a posterior root, the latter showing an oval enlargement called the ganglion. These nerve roots join to form plexuses. Thus we have:

1. The *cervical plexus,* formed by the anterior divisions of the upper 4 cervical nerves
2. The *brachial plexus,* which is formed by the anterior divisions of the cervical nerves 5 to 8, and the first thoracic nerve
3. The *lumbar plexus,* formed by the anterior divisions of the lumbar nerves 1, 2, 3 and the greater part of lumbar 4
4. The *sacral plexus,* formed by the roots of lumbars 4 and 5, and sacrals 1, 2 and part of 3
5. The *coccygeal plexus,* formed by the sacral nerves 3 to 5

In addition to the 12 pairs of intercostal nerves, some of the most significant nerves are the iliohypogastric nerve arising from the first lumbar root; the ilio-inguinal nerve arising also from the first and the second lumbar roots; the sciatic nerve, which is the main nerve of the sacral plexus; and the coccygeal nerve, which also receives filaments from the fourth and the fifth sacral nerves.

Exit of Spinal Nerves

As the anterior root of the spinal cord emerges from the anterior and lateral gray columns, it traverses the surrounding membranes of pia, arachnoid and dura. The posterior root, which is attached to the posterolateral portion of the spinal cord, originates from 2 bundles of fibers in the spinal ganglion. Both anterior and posterior roots pierce the dura separately as they make their exit

through their respective intervertebral foramina. As a rule, the posterior root is thicker and larger than the anterior root. They are enclosed in a common dural sheath just beyond the spinal ganglion where they become the spinal nerve and are surrounded by epineurium.

The spinal ganglia, which lie at the outer portion of the intervertebral foramina, are oval-shaped and vary in size corresponding to their nerve roots.

The spinal nerves lie horizontally in the cervical region, but below these segments the spinal nerves assume an increasingly oblique and downward direction as they approach the lumbar region where they are almost vertical, forming the cauda equina. At the lower thoracic level there is a difference of 2 vertebral segments between the origin of the spinal nerve and the level of exit.

From each sympathetic trunk ganglion, which lies on the posterolateral surface of the vertebral body, a branch (gray ramus communicans), joins the adjacent spinal nerve.

Efferent, preganglionic sympathetic fibers (white ramus communicans), which originate in the lateral columns, pass along with the anterior root to the corresponding sympathetic ganglion or along its trunk to sympathetic plexus.

Shortly after emerging from the intervertebral foramen each spinal nerve turns back through the same foramen to supply the spinal cord membranes, blood vessels, intervertebral ligaments and joint surfaces.

The spinal nerve then divides into 2 branches, each with fibers from both roots.

1. *Anterior division* supplies the anterior and the lateral portions of the trunk and the limbs. In the thoracic region it spans the space between the pleura and the intercostal membranes, runs below the lower rib margin and supplies the intercostal muscles and adjacent skin. In the cervical and lumbar regions the anterior divisions form plexuses.

2. *Posterior division* is directed backward shortly beyond the formation of the spinal nerve. Its medial branch supplies the multifides, the longissimus, the semispinalis and the trapezius muscles, then proceeds along the spinous process and supplies the skin. Its lateral branch traverses the longissimus muscle and supplies the intercostal muscle and adjacent skin.

In the lumbar region the medial branches of the posterior division hug the articular processes of the vertebrae and end in the multifides, and the lateral branches supply the group of sacrospinalis muscles, adjacent fascia and skin.

Sections Through the Spinal Cord

Cross sections of the spinal cord at various levels show considerable variation in size and shape. The proportion of gray to white matter also varies and is much greater in the cervical and the lumbar regions and greatest in the conus medullaris. The anterior and the posterior gray columns in the thoracic region are equally thin, but in the cervical region the anterior gray columns are larger; in the lumbar region and below, both gray columns are about equally wide and in much greater proportion to the white matter.

Through the entire length of the spinal cord runs the central canal, which is lined with ciliated ependymal cells. Superiorly, it opens into the fourth ventricle, and inferiorly extends into the filum terminale. The horizontal gray matter which joins the gray columns surrounds the central canal and is divided by it into anterior and posterior gray commissures.

Tracts

Through the spinal cord run fibers carrying impulses to and from various portions of the brain. These fibers group themselves into tracts. Only those of known clinical importance will be described.

Funiculus Gracilus (Goll) and Funiculus Cuneatus (Burdach). These carry muscle and joint sensations and lie between the posterior median and the posterolateral sulcus. In the cervical and the thoracic regions these tracts are separated by a septum at the lower portion of which is found the Comma Tract (Schultze).

Lateral Spinothalamic Tract. This tract mediates pain and temperature sensation. It arises in the posterior column, crosses to the opposite side in the anterior commissure and ascends in the lateral funiculus to the thalamus.

Ventral Spinothalamic Tract. This tract transmits impulses of touch. It also arises in the posterior column, crosses in the anterior

commissure to the opposite side and ascends in the anterior funiculus to the thalamus.

Dorsal Spinocerebellar Tract. This tract transmits impulses from leg muscles and trunk between the sixth cervical and the second lumbar segments. It is located on the lateral surface ventral to the posterolateral sulcus and ascends to the cerebellum via the restiform body.

Ventral Spinocerebellar Tract (Gower's). This tract carries impulses to the cerebellum via the medulla, the pons and the anterior medullary velum. It lies at the periphery on the ventrolateral aspect of the cord.

Spinotectal Tract. This tract arises from cells in the posterior gray column, crosses over and ascends in the lateral funiculus and ends in the corpora quadrigemina.

Rubrospinal Tract. This tract carries impulses for cerebellar reflexes. It arises in the red nucleus, crosses over and descends near the center of the lateral funiculus.

Lateral Pyramidal Tract. This tract carries impulses to the primary motor neurone. It arises from large cells in the precentral gyrus and, after decussation in the medulla, enters the lateral funiculus lying between the dorsal spinocerebellar tract and the lateral funiculus.

Direct Pyramidal Tract. This tract is small; it arises from cells in the central motor area, passes down the same side close to the anterior median fissure, then crosses in the anterior commissure to the opposite side and at various levels ends by synapes with the anterior horn cells.

Tectospinal Tract. This tract mediates optic and auditory reflexes. It arises in the superior colliculi, crosses and then descends in the anterior funiculus to end in the motor cells of the anterior column.

Spinal Membranes and Nerve Roots

The spinal cord is enveloped by membranes which are a direct continuation of those surrounding the brain.

Dura Mater. This extends direct from the cranial dura, beginning at the foramen magnum and continuing as far down as the second sacral vertebra. This membrane adheres anteriorly to the posterior longitudinal ligament and corresponds in shape to the enlargements of the spinal cord. The dura also invests the spinal nerves as they leave the lateral margins of the spinal cord to their points of exit.

Subdural Space. This is a potential space containing a minute quantity of fluid and is found between the arachnoid and the dura.

Arachnoid and Subarachnoid. These consist of a delicate meshwork of mesothelial cells with spaces filled with cerebrospinal fluid. These membranes also envelop the spinal nerves to their intervertebral foramina.

Pia Mater. This delicate fibrous layer intimately invests the spinal cord. At its lateral margins the pia forms denser pointed prolongations to the inner dural surface. These are spaced equally between the nerve roots, and also separate the anterior from the posterior spinal roots.

Although there are no visible demarcations, the spinal cord is said to be made up of segments of varying lengths. These segments correspond approximately to the attachments of a pair of spinal nerves. The widest segments are in the midthoracic region.

On cross section of the spinal cord one can see the *white matter* on the periphery, which is made up of medullated nerve fibers held together by neuroglia. In the central portion of the spinal cord we find the *gray matter*, which has the form of the letter "H." This consists of numerous nerve cells and nonmedullated nerve fibers, held together by neuroglia.

Spinal Nerves. These consist of an anterior spinal root and a posterior spinal root with its ganglion. These are attached to the corresponding gray matter of the spinal cord.

Anterior Root. This root arises from nerve cells in the anterior and lateral columns of the gray matter, and as they pass through the white matter they become medullated. They leave the spinal cord in 2 or 3 irregular rows (fila) to efferent pathways.

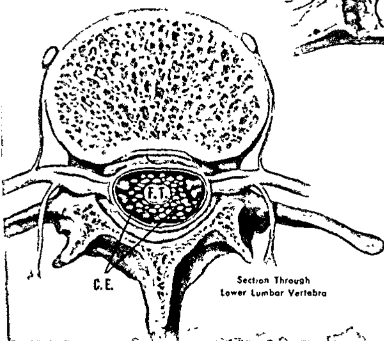
Posterior Root. This root arises from the medial afferent fibers of the spinal ganglion and reaches the posterolateral sulcus in the form of 6 or 8 fasciculi (fila).

Spinal Ganglia. These ganglia are enveloped by the continuation of the dural sheath and contain irregularly spherical cells. These cells give off a unipolar coiled axone which divides into a medial and a lateral portion. The former is directed toward the spinal cord and becomes the posterior root, while the latter is directed peripherally to sensory end organs of muscles, joints, skin and viscera.

Anterior Median Fissure. This fissure is lined by an overlapping fold of pia and dips

Section Through Thoracic Vertebra

S.T.
P.P.
L.
R.C.
D.M.
E-D.S.
A.
A.R.
S.C.
S.G.
A.Ra.
P.R.
P.Ra.
S-A.S.
S.
L.
R.
R.
S.P.
F. Netter M.D.



A. — Arachnoid
A R. — Anterior Root of Spinal Nerve
A Ra. — Anterior Ramus of Thoracic Nerve (Intercostal Nerve)
C E. — Cauda Equina
D M. — Dura Mater
E D S — Epidural Space
F T. — Filum Terminale
L. — Lamina
L. — Lung
P P. — Parietal Pleura
P Ra — Posterior Ramus of Thoracic Nerve
P R. — Posterior Root of Spinal Nerve
R. — Rib
R C — Rami Communicantes
S. — Subarachnoid Septum
S-A S. — Subarachnoid Space
S C. — Spinal Cord
S G — Spinal Ganglion
S P — Spinous Process
S T — Sympathetic Trunk
V B — Vertebral Body

PLATE 15. Exit of spinal nerves. (Drawing by F. Netter *in* The central nervous system, Ciba Clinical Symposia 1:187, No 6)

DORSAL

VENTRAL

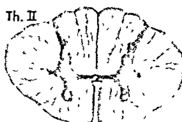
C T. — Comma Tract
D P. — Direct Pyramidal Tract
D S -C — Dorsal Spinocerebellar Tract
F C. — Funiculus Cuneatus (Burdach)
F G. — Funiculus Gracilis (Goll)
L P. — Lateral Pyramidal Tract
L S -T. — Lateral Spinothalamic Tract
O B. — Oval Bundle
R S — Rubrospinal Tract
S T — Spinotectal Tract
T S. — Tectospinal Tract
V.S -C. — Ventral Spinocerebellar Tract (Gower)
V S -T — Ventral Spinothalamic Tract

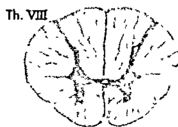
Schematic section through spinal cord, showing on left the tracts of greatest clinical importance and, on right, other tracts and landmarks as well.

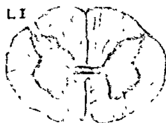
(RED indicates tracts from brain to cord, BLUE, from cord to brain)

A. — Apex of Posterior Column
A G C. — Anterior Gray Commissure
A.W.C — Anterior White Commissure
B S. — Bulbospinal Tract (Helwig s Bundle)
C. — Cervix of Posterior Column
F P — Fasciculus Proprius
F R. — Formatio Reticularis
M Z — Marginal Zone
N D. — Nucleus Dorsalis
P.G.C. — Posterior Gray Commissure
S G — Substancia Gelatinosa
S-M F — Sulco-Marginal Fasciculus
V S — Vestibulospinal Tract

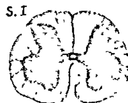


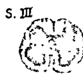












Representative sections through cord at various levels

F. Netter M.D.

PLATE 16. Sections through the spinal cord. (Drawing by F. Netter *in* The central nervous system, Ciba Clinical Symposia 1:188, No. 6)

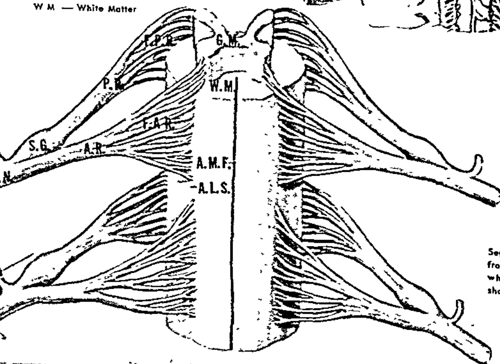
Segment of spinal cord viewed from behind, with portions of dura mater and arachnoid removed.

D.M.
A.
A.S.
P.R.
D.L.
P.

A. — Arachnoid
D L — Dentate Ligament
D M. — Dura Mater
P. — Pia Mater Overlying Spinal Cord
P R. — Posterior Root
S-A S. — Subarachnoid Septum

A L S — Anterior Lateral Sulcus
A M.F — Anterior Median Fissure
A R — Anterior Root
F.A R. — Fila of Anterior Root
F P R — Fila of Posterior Root
G M. — Gray Matter
S G — Spinal Ganglion
S N. — Spinal Nerve
W M — White Matter

F. Netter M.D.



Segment of spinal cord, viewed from in front with portion of white matter removed and showing origin of spinal nerves.

PLATE 17. Spinal membranes and nerve roots. (Drawing by F. Netter *in* The central nervous system, Ciba Clinical Symposia 1:189, No. 6)

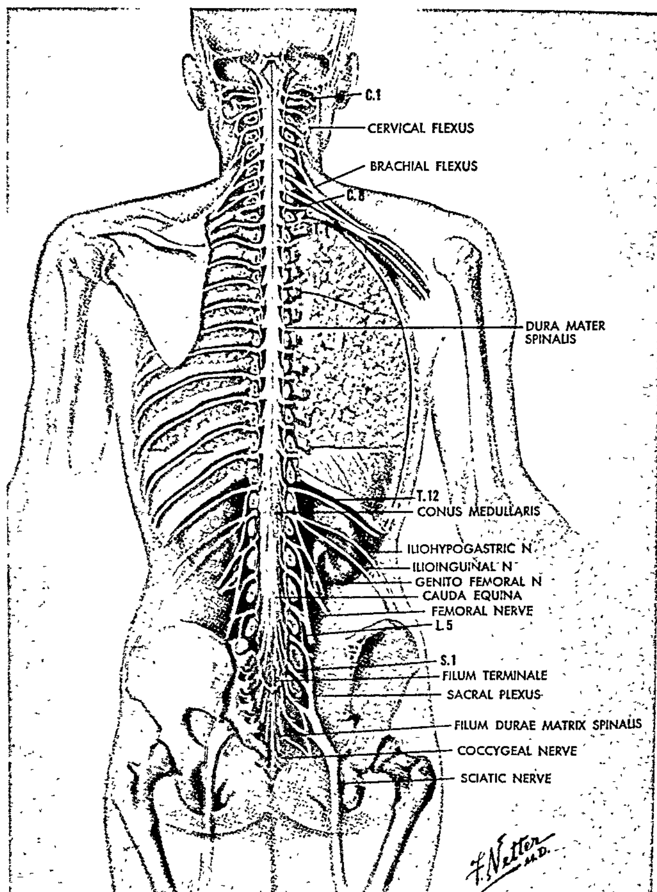


FIG. 134. The spinal cord in situ. (Drawing by F. Netter *in* The Central Nervous System, Clinical Symposia 1:186, No. 6)

veins, on either side of the spinal cord, and they all empty into the intervertebral veins. However, those near the foramen magnum empty into the inferior petrosal sinus of cerebellar veins.

Dermal Segmentation

Sensation from the outside world reaches consciousness through sensory impulses. Most of these are carried by afferent nerve fibers to the spinal cord and up to the brain.

The nerve fibers which carry sensation of pain, temperature, touch, vibration, position sense and other discriminatory sensibilities have their cells of origin in the spinal ganglia from which arise also fibers to make up the dorsal root.

The fibers that carry the impulses of sensation from the skin, the muscles and the joints are arranged in segments which in simplest form are found in the thoracic region as broad bands. In the upper and the lower limbs the sensory arrangement is more complicated but follows a vertical pattern in each limb.

It has been found that if a dorsal root is sectioned, complete anesthesia of the involved dermal segment does not follow because of an overlapping of sensation by the nerves above and below the affected dermal segment. It has been further established that each sensory nerve carries impulses not only from its own dermal segment but also from the ones above and below. This overlapping of cutaneous sensation is known as metamerism.

If one is familiar with the cutaneous distribution of various nerve roots it is possible to localize with great accuracy the site and the level of any pathologic disturbance.

A chart outlining the exact sensory dermal segments is a good reference, but it is valuable to remember some surface landmarks which will serve as a general guide to localization.

1. The clavicle is supplied by Cervical 3 sensory root.

2. The deltoid is supplied by Cervical 5 sensory root.

3. The nipple area is supplied by Thoracic 4 sensory root.

4. The intercostal margin is supplied by Thoracic 7 sensory root.

5. The umbilicus is supplied by Thoracic 10 sensory root

6. The groin region is supplied by Thoracic 12 sensory root.

7. The lateral aspect of the arm is supplied by Cervical 6, 7 and 8 sensory roots.

8. The inner aspect of the arm is supplied by Thoracic 1 and 2 sensory roots.

9. The inner and anterior surface of the leg is supplied from above down by Lumbar 1, 2, 3 and 4 sensory roots.

10. The outer and posterior surfaces of the legs are supplied by Lumbar 5 and Sacral 1 and 2 sensory roots.

11. The perineum is supplied by Sacral 3, 4 and 5 sensory roots.

NEUROLOGIC DIAGNOSIS[2]

Diagnosis of orthopaedic conditions requires a basic understanding of neurology. A simple neurologic examination ordinarily suffices to differentiate the majority of neurologic disorders.

Motor Function

Disturbance of muscle power varies from *paresis* (weakness) and *paralysis* (complete loss) to *hyperkinesia* (increased muscular movements). The patient may be able to move a muscle, yet paresis may be demonstrated by inability to perform a movement against resistance. *Monoplegia* defines paralysis of a single extremity; *hemiplegia* is paralysis of a unilateral half of the body; *diplegia or brachial paraplegia* is paralysis of both upper extremities; and *paraplegia* is paralysis of both lower extremities. The paralysis is due either to an upper or a lower motor neuron lesion.

The upper motor pyramidal cells of the cerebral precentral cortex send fibers through the corticobulbar and the corticospinal tracts to the motor cells of the cranial nerves and of the anterior horns of the spinal cord on the opposite side. These tracts undergo partial decussation at the caudal end of the medulla before continuing distally into the spinal cord. The major portion of the fibers cross to the opposite side, forming the lateral corticospinal tract; the smaller uncrossed portion continues downward as the ventral corticospinal tract.

[2] Davis, L.: Neurologic Diagnosis, Philadelphia, Saunders, 1923

The lower motor neurons are also under the influence of various other motor centers in the brain, located chiefly in the basal ganglia and the cerebellum.

Lesions

Upper motor neuron lesions produce a spastic paralysis characterized by increased muscle tone, increased deep reflexes, diminished or absent superficial reflexes, and demonstrable pathologic reflexes such as the Babinski, the Oppenheim, the Gordon and the Chaddock. These findings are explained by the removal of the inhibitory impulses of the cerebral centers.

A lower motor neuron lesion is characterized by a flaccid paralysis (loss of muscle tone), absent deep reflexes, muscle atrophy and the reaction of degeneration. This type of paralysis may be produced by disease or injury to the anterior horn cells, the anterior roots, the peripheral nerves, the nerve plexuses, or the cauda equina.

Hyperkinesia is a condition of excessive, involuntary, purposeless movements.

Tremors are rhythmic, oscillating movements affecting all or groups of muscles.

Intention tremors are characteristic of multiple sclerosis.

Tonic spasms are prolonged, intense, muscular contractions.

Clonic spasms are rapid, repeated contractions of muscles.

A cramp is a tonic spasm localized to one muscle. Persisting spasm of a muscle eventually leads to its contracture. Continued spasm of a group of muscles overcoming their antagonists may cause joint contracture.

Choreiform movements are quick, un-co-ordinated, irregular and arrhythmic; they are characteristic of chorea which commonly follows rheumatic fever.

Athetosis, which is due to basal ganglia damage and often associated with hemiplegia, is characterized by a recurring series of slow, vermicular, "pill-rolling" movements of the hands.

Myotonia is a condition of increased muscle tonus which is brought on by emotion and attempts at movement.

Lesions of the corpus striatum are commonly associated with cerebral palsy. They are characterized by athetochoreic movements, rigidity, tremor, loss of associate movements, and masked facies.

Synergic movements are governed by the cerebellum.

Adiodokokinesis is the inability to accomplish synergic movements, e.g., the patient is unable to perform rapidly and alternatingly supination and pronation with both hands at the same time.

Hypotonia is a decrease of muscle tonus associated with muscle atrophy. Hypotonia also arises when lesions interrupt transmission of deep sensation, e.g., in tabes dorsalis.

Gait. Complicated co-ordinated movements are examined by observing the manner of walking. Paresis will produce a slow guarded, short-stepped, shuffling gait. Paralysis of the anterior tibial muscles, especially by an anterior horn or peripheral nerve lesion, causes a drop foot and produces a *steppage* gait. To avoid tripping over the plantar-flexed foot, the extremity is advanced with knee and hip hyperflexed. With *spasticity,* the legs are advanced slowly with shortened steps and the toes scraping the ground. Adductor tightness produces a scissors gait by which the legs are alternately crossed. In the *ataxic or tabetic gait,* because of absence of deep position sense, the patient must constantly observe the placing of his feet; the hip is hyperflexed and externally rotated, and the forefoot is strongly dorsiflexed before being thrown down with the heel striking the ground first. The patient is unable to stand with the eyes closed. In contrast, the cerebellar ataxic is not aided by visual assistance. The gait is stumbling, drunken, swaying from side to side, and there is a tendency to fall toward the side of the lesion.

Muscle co-ordination in the lower extremity may be tested by having the patient, with his eyes closed, place the heel of one foot upon various points on the opposite leg. The upper extremities are tested by asking the patient to touch the tip of his nose with the end of the index finger.

Electrical Reactions

The reaction of degeneration, characteristic of a lower motor neuron lesion, appears 2 weeks after the lesion. Normal muscle re-

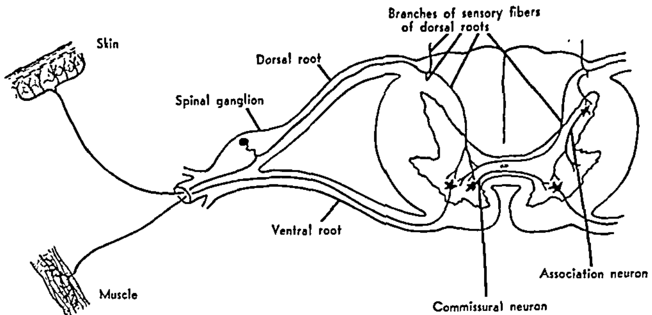


FIG. 135. Elements of the spinal reflex arc. (After Ranson)

sponds to both faradic and galvanic currents. Faradic current causes a tonic contraction which persists as long as the current flows through the muscle. Galvanic current causes a single contraction with the cathode only, the negative closing contraction. With the reaction of degeneration, the muscle fails to react normally to electrical stimulation:

1. Faradic current to nerve or muscle fails to cause a contraction.

2. Galvanic current applied to the nerve fails to cause a contraction.

3. Galvanic current applied to the muscle causes a sluggish contraction. A reversal of response is characteristic; the positive (anodal) closing contraction (A C.C.) is greater than the negative (cathodal) closing contraction (C.C.C.).

REFLEXES

The simplest spinal reflex consists of a primary sensory and motor neuron and a synapse in the anterior gray matter of the spinal cord. It is composed of (1) a receptor, or peripheral sensory nerve ending; (2) an afferent conductor; (3) a synaptic center; (4) an efferent conductor; (5) and the effector mechanism or muscle fibers. One or more intermediate neurons are interposed between the primary neurons. The latter may remain localized to one side of the cord (association neurons), pass to the opposite side of the cord (commissural neurons), or extend proximally or distally to complete an intersegmental reflex arc. Each reflex is contained within a definite segment of the cord.

Deep Reflexes. Reflexes requiring stimulation of the tendons, e.g., patellar, Achilles, biceps and triceps, are termed deep reflexes.

Superficial Reflexes. Others requiring cutaneous stimulation, e.g., abdominal and cremasteric, comprise the superficial reflexes. Destruction of either limb of the reflex arc or the spinal cord segment abolishes the reflex response. If the lesion extends across the entire spinal cord segment, elimination of inhibitory impulses from cerebral centers causes an exaggeration of deep reflexes below the level of the lesion. Superficial reflex arcs involve the cerebral cortex, thereby explaining their absence in upper motor neuron lesions.

Pathologic Reflexes. Destruction of the upper motor neurons or pyramidal tracts is indicated by pathologic reflexes. The Babinski phenomenon is elicited by stroking the plantar surface of the foot. Normally, all the toes flex plantarward. A pathologic response is a dorsiflexion of the large toe. An exaggerated deep reflex is only of significance if associated with absent superficial reflexes, pathologic reflexes and sustained clonus. Sphincter disturbances are often present, manifest by difficulty in

starting the urinary stream, urinary retention or incontinence.

Argyll Robertson Pupil. The pupil of the eye contracts when exposed to light and when accommodating for a near object. When the pupil reflexly contracts to accommodation but not to light, it is the Argyll Robertson pupil *characteristic of central nervous system lues.*

Reflex Centers. The following are the centers of various reflexes of importance to the orthopaedic surgeon:

1 DEEP REFLEXES	
Biceps	CV
Triceps	CVI
Radial	CVIII
Ulnar	TI
Patellar	LII-III
Achilles	LV-SI
2. SUPERFICIAL REFLEXES	
Upper abdominal	TVIII-X
Lower abdominal	TX-XII
Cremaster	LII
3 SPHINTERIC REFLEXES	
Bladder	SIII-IV
Anus	SIII-IV

Mass Reflexes. In normal man, the motor response to an afferent impulse is localized and specific. In lower vertebrates the response, usually a flexor spasm, is more widespread and constitutes a protective reflex. When, in the human being, the spinal cord is completely transected, the distal portion temporarily loses and then regains its reflex excitability. The reflex response is now primitive Specificity of response is lost, and stimulation occasions a widespread motor reaction. For example, stroking the plantar surface of the foot produces flexion at the hip and the knee, dorsiflexion of the foot, and emptying the bladder. This is the "mass reflex" characteristic of complete interruption in continuity of the spinal cord. The center for this reflex is located low down in the spinal cord and is independent of cerebral control.

SENSATION

Objective evidence of sensory loss defines the site of the lesion and should be compared with normal areas. Changes of sensation form an accurate indication of improvement or progression. *Light touch* is determined by stroking the skin with a wisp of cotton. The hair upon the skin produces sensation other than light touch and should first be removed. *Pressure touch* is tested by use of a blunt instrument. Pricking the skin with a needle elicits *superficial pain. Temperature* sensation is determined by applying a test tube filled with hot water and another with cold water. *Vibration* sense is tested with a tuning fork.

Epicritic or discrimination sensibility is the ability to discriminate between two points. When this sensation is reduced, the points of a compass may have to be widely separated before the stimulus can be recognized as dual. *Position* is tested by placing a part of an extremity such as the large toe in a certain attitude and asking the blindfolded patient to describe the position. *Stereognosis,* the sensation of size, shape and form, the center for which exists in the parietal lobe, is determined by placing familiar objects in the patient's hand. Disease of the posterior columns of the cord produces loss of muscle and joint sensibility in the hands and, therefore, a loss of stereognosis.

Sensations conducted through peripheral nerves consist of: (1) *deep sensation,* the ability to discern pressure, position and movement; (2) *protopathic sensation,* the recognition of painful stimuli and the distinction between extremes of hot and cold; (3) *epicritic sensation,* the ability to discriminate between two points and to distinguish between finer grades of temperature.

Severance of a cutaneous nerve produces a loss of all forms of superficial sensation, touch, pinprick, 2-point discrimination, and distinction between hot and cold. The sense of deep position, pressure and vibration is preserved. Epicritic sensory loss is well defined. Protopathic sensory loss after division of a peripheral nerve is smaller because of enormous overlapping in innervation from several nerves Therefore, in determining peripheral sensory loss, testing superficial touch with a wisp of cotton is more accurate than testing pain perception with a pin. Destruction of nerves closer to the spinal cord increases the extent of loss to painful stimuli.

Afferent sensory fibers pass through the dorsal root to enter the spinal cord. Each fiber passes to the posterior gray column where it

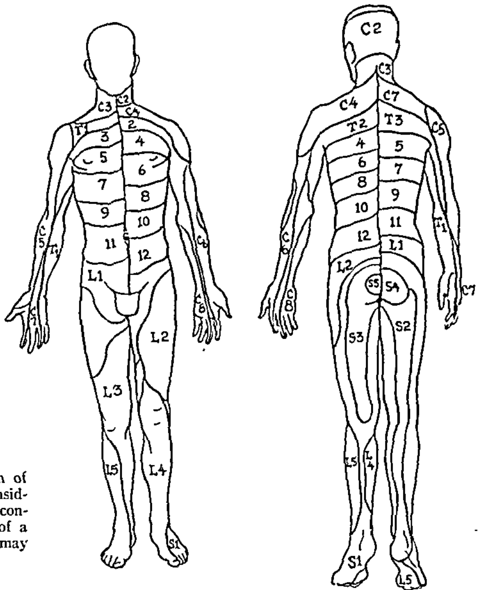


Fig. 136. Distribution of spinal dermatomes. Considerable overlap occurs; consequently, involvement of a single spinal segment may not be evident.

divides into a long ascending and a short descending branch. Thus synapses are made not only with neurons at the same level but at other levels as high as the medulla. Through relays, afferent impulses reach the cerebral cortex and the cerebellum.

The impulses of movement, position and vibration pass upward within the cord on the same side as their point of entry. Impulses of touch, pain and temperature cross to the opposite side of the cord before ascending

A transverse division of the spinal cord causes loss of sensation below the level of the lesion. At the upper level of sensory loss there exists a band of hyperesthesia due to sensory root irritation at the level of the lesion.

Sensory supply to the body is made up of a regularly spaced series of dermatones which correspond to spinal cord segments. If one imagines the body in the quadruped position and then intersects the body at regular intervals, beginning at the neck and ending at the coccyx, the segmental nerve distribution will be apparent. In this position the thumbs and the large toes are in a more advanced position than the small finger and toe. Therefore, the radial side of the upper extremity is represented by a higher segmental level of the spinal cord than the ulnar side; the medial side of the thigh and the leg are of a higher segmental level than the external side of the lower extremity. By finding the sensory loss in a specific dermatone, the level of the spinal cord lesion is localized.

The viscera are generally lacking in sensory fibers. Disease in a viscus supplied by a certain spinal cord segment will produce referred pain in the cutaneous distribution of that seg-

CHARACTERISTIC VOLTAGES OF NORMAL & ABNORMAL SKELETAL MUSCLE

AT REST

Normal	Denervated	Partially Denervated
Complete Electrical Silence	Fibrillation Voltages	Fibrillation & or Fasciculation Voltages

DURING VOLUNTARY EFFORT

Simple Motor Unit Voltages	Fibrillation Voltages	Simple & or Complex Motor Unit Voltages

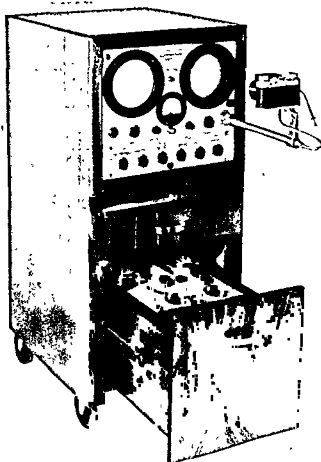


Fig. 137. Diagnostic electromyography. In the illustration above, normal and abnormal voltages are recorded on *graphs. Below is shown the cathode ray* oscillograph and the camera position for making a permanent record. (Golseth, J. G: Diagnostic contributions of the electromyogram, California Med. 73: 355)

ment. Thus, gallbladder disease causes interscapular pain; subdiaphragmatic disease causes pain in the shoulder.

The type of pain should be noted. Tabes dorsalis causes lightning pains, neuritis causes a burning pain, arthritis an aching pain and sense of stiffness, and spinal disease a girdle pain.

The Autonomic Nervous System

From various centers in the central nervous system, fibers passing through spinal nerves pass via rami communicantes to ganglia; the latter in turn innervate the viscera, the glands, the heart, the blood vessels and the smooth muscles. The sympathetic trunk, on each side of the body [illegible] l aspect

of the spinal column from the level of the second cervical vertebra to the coccyx. It is composed of 21 to 22 ganglia, 3 cervical (*superior, middle, inferior*), 10 or 11 thoracic, 4 lumbar and 4 sacral. Gray and white rami communicantes connect the sympathetic trunk with the spinal nerves. The gray rami are composed of unmyelinated fibers originating in the sympathetic ganglia and distributed through peripheral nerves to blood vessels (vasomotor), sweat glands (secretory), and smooth muscles of hair follicles (pilomotor). The white rami consist of myelinated fibers which originate from cells in the intermediolateral column of the spinal cord and form preganglionic fibers which synapse with ganglionic cells. The abdominal portion of the sympathetic trunk supplying the lower extremity and the inferior, or stellate, ganglion of the cervical trunk supplying the upper extremity are of importance to the orthopaedic surgeon. A great portion of sympathetic fibers innervating blood vessels pass distally in the vessel walls and are independent of peripheral nerves.

Preganglionic fibers which originate in the brain and the sacral region run directly to ganglia and their plexuses which lie distally near the innervated organ. These react similarly to pilocarpine and are termed "parasympathetic." Preganglionic fibers originating in the thoracolumbar region of the cord pass to ganglia in the sympathetic chain which lies next to the spine and at a distance from the innervated part. These react to epinephrine and are termed "sympathetic."

Most structures are innervated from both sympathetic and parasympathetic divisions, the action of such supply counteracting each other. Thus, a blood vessel will undergo spasm from action of adrenergic drugs but relaxes on administration of cholinergic drugs.

Trophic Innervation

A trophic center in the anterior horn is concerned with nutrition of all parts of the body. A lesion affecting this center or its peripheral nerve results in trophic changes. An acute irritative lesion produces herpes zoster. A chronic irritative lesion produces a thickened, rough, scaly skin. A destructive [illegible] come ridged and brittle. The muscles become atrophic, and bones undergo osteoporosis. Severe, painless, destruction of joints associated with tabes dorsalis and syringomyelia is thought to be trophic in origin.

ELECTROMYOGRAPHY[3]

When muscle fibers contract, they generate an electric current which can be seen and heard on the electromyograph. A needle electrode is inserted into the muscle, and the minute voltages are amplified, converted into visible patterns by means of a cathode ray oscillograph and transformed into sound energy causing characteristic sounds over a loud speaker. The screen patterns can be photographed and a permanent record obtained. The electromyogram is a useful diagnostic adjunct in interpreting the site of disorders in the central nervous system or the musculature.

A relaxed normal voluntary muscle generates no current, a straight line appearing on the oscillograph screen. A normal voluntary muscle, when contracting by voluntary or reflex effort, will generate "normal motor unit voltages," the screen displaying simple waves which are monophasic, diphasic, or triphasic.

A motor unit is defined as a functional unit consisting of one anterior horn cell, its axon and 100 to 150 muscle fibers which are innervated by the axon.[4] When the axon of this motor unit is excited, all of the muscle fibers appear to contract synchronously, resulting in the spread of a contraction wave which generates the characteristic "normal motor unit voltage." Normal motor unit voltages range from 100 to 2,000 microvolts and can be elicited from any area of the contracting muscle. The duration of each wave is from 3 to 10 milliseconds, and their frequency varies from 5 to 30 per second, depending upon the force of contraction. Because of their simple wave form and relatively long duration, the normal motor unit voltages produce a very characteristic thumping noise in the loud speaker.

Individual muscle fibers, when denervated for a period of time, will twitch in a rhythmic

[3] Golseth, J. G.: Diagnostic contributions of the electromyogram. [illegible]

manner. This involuntary contraction is termed "denervation fibrillation." True denervation fibrillation cannot be observed clinically through the intact skin inasmuch as it represents contraction of the individual muscle fibers. Therefore, to detect its presence electromyography is employed. The voltages generated by fibrillating denervated muscle fibers have a short magnitude ranging from 5 to 100 microvolts and are termed "denervation fibrillation voltages." Their form is diphasic, and their frequency varies from 2 to 30 per second. Each wave has an extremely short duration of 1 to 2 milliseconds, which is responsible for the characteristic clicking sound.

When regeneration of the motor nerve takes place, the reactivated muscle generates motor unit voltages which produce complex highly polyphasic waves. These polyphasic waves are also characteristic of muscles affected by demyelinating diseases of the anterior horns, namely, poliomyelitis, amyotrophic lateral sclerosis and progressive muscular atrophy. Therefore, these abnormal electromyographic waves represent early nerve degeneration or early nerve regeneration. The complex or highly polyphasic wave is apparently caused by the muscle fibers contracting asynchronously in contrast with the normal muscle in which the fibers contract synchronously. The complex motor unit voltages range in magnitude from about 1,000 to 1,500 microvolts, vary in frequency from 2 to 30 per second, and the duration of each wave is from 5 to 15 milliseconds Irregularity of these motor unit voltages give rise to a very rough-sounding noise

Because denervation fibrillation takes place only after Wallerian degeneration of the nerve supply has taken place, it is a valuable objective sign of lower motor neuron disease. Its presence differentiates between upper and lower motor neuron disease, myopathy and atrophy, and functional and organic paralyses

Denervation fibrillation can be used for localization of a lower motor neuron lesion For example, compression of a nerve root can be identified by fibrillation in a group of muscles constituting the innervated myotome When the *fifth cervical root* is involved, fibrillation is present in the sacrospinalis between the 4th and the 5th cervical vertebrae (posterior primary division), rhomboids, supraspinatus, infraspinatus, deltoid, biceps and brachioradialis (anterior primary division). In disk protrusion at the lumbosacral level, the *first sacral nerve root* compression will cause fibrillation and abnormal motor units in the gastrocnemius, the hamstrings, the gluteus maximus (anterior primary division) and the erector spinalis between the 1st and the 2nd sacral vertebrae. Electromyographic changes are most pronounced when root injury is severe and prolonged. In diagnosing a root compression syndrome, electromyography must be used in conjunction with myelography and a neurologic examination. When a *peripheral nerve*, such as the median, is involved, denervation fibrillation will be observed only in those muscles supplied by that nerve.

In *muscular dystrophy*, instead of the normal simple motor unit voltages on voluntary effort, the waves are of lesser magnitude (up to 150 microvolts) and shorter (1 to 2 milliseconds). When the patient is completely relaxed, no fibrillation is noted. In *myotonia*, on voluntary effort a burst of impulses may continue for 30 to 50 seconds, but there is no fibrillation. In *myasthenia gravis*, there is first a burst of impulses which rapidly diminish in size, amplitude and frequency. Characteristically in myasthenia gravis, Prostigmine causes temporary restoration of normal motor unit voltages.[5]

Although denervation fibrillation indicates that some of the muscle fibers are without innervation, it also indicates that these fibers are still capable of contractility. Therefore, the nutrition of these muscles may be preserved theoretically by interrupted galvanic stimulation, massage and heat; irreversible fibrotic changes are thereby prevented until, as in the case of a peripheral nerve lesion, regeneration takes place.

LOCALIZATION OF SPINAL CORD LESION[6]

Basic Anatomy

The pyramidal, or motor, tracts which have already crossed above the spinal cord descend in the lateral columns of white matter. Anterior and lateral to these fibers are the lateral

[5] Marinacci, A A . The value of electromyography in neurology, California Med 80 314, 1954

[6] Davis, L Neurological Surgery, ed 2, Philadelphia, Lea & Febiger, 1942

spinothalamic tracts which carry impulses of pain and temperature upward from the opposite side of the body. The posterior white columns transmit upward both deep (joint, muscle, bone) and superficial tactile sensations uncrossed. The following are examples:

Types of Lesions

Lower Motor Neuron Lesion. In this lesion limbs are flaccid, deep reflexes are absent, and muscles are atrophied. This lesion may be in the anterior horn, the anterior spinal root, or a peripheral nerve. If sensory changes are absent, the peripheral nerve is excluded, and anterior poliomyelitis is the commonest cause.

One must remember that a sudden traumatic lesion of the spinal cord produces at first a flaccid paralysis below the level of the lesion, but eventually spastic paralysis develops.

Upper Motor Neuron Lesion. This condition is characterized by increased muscle tone and spasticity, hyperreflexia, clonus, absent superficial reflexes and positive pathologic reflexes. Bilateral spastic paralysis is due most commonly to cerebral palsy.

Combined Upper and Lower Motor Neuron Lesion. Spastic paralysis in the lower extremities and flaccid paralysis in the upper extremities with a reaction of degeneration and no sensory loss indicate a combined lesion of the anterior horns and the corticospinal tracts. The disease is amyotrophic lateral sclerosis. When this combination is associated with loss of pain and temperature sense, while tactile sense is preserved, the cause is syringomyelia or an intramedullary tumor. The central lesion destroys the centrally crossing fibers of pain and temperature while tactile fibers ascend in the posterior columns unharmed.

Lesion of Posterior Spinal Root. This lesion involves absence of deep reflexes, loss of all sensation, and spontaneous lightning pains. Loss of joint and muscle sense results in ataxia. The disease is due to tabes dorsalis (locomotor ataxia). The posterior columns undergo secondary degeneration.

Combined Lesion of Posterior Columns and Pyramidal Tracts. In addition to loss of joint and muscle sense and an ataxia, a spastic weakness with hyperreflexia is present Subacute combined degeneration is due to pernicious anemia. A high color index and achlorhydria are confirmatory findings.

Transverse Lesion of the Spinal Cord. At the corresponding segmental level, the deep reflex is absent; flaccid paralysis, muscle atrophy and reaction of degeneration are present. Below this level, one finds spastic paralysis, hyperreflexia, absent superficial reflexes, pathologic reflexes, complete sensory loss, sphincteric constriction with urinary retention. The cause of the pathology includes myelitis, tumor, fracture-dislocation of the spine, thrombosis, hemorrhage, abscess and vertebral disease. When the onset is sudden, vascular or traumatic lesions are probable. Tumors and infections cause slowly progressive symptoms.

SPINAL CORD TUMORS

To the orthopaedic surgeon the identification of a spinal cord tumor is important as a source of pain about the spine or of pain referred to other regions. Symptoms about the chest and the abdomen may cause confusion with visceral disease. Especially important is the differentiation from other causes of sciatica, particularly a ruptured intervertebral disk.

Pathology[7]

These are *intramedullary* tumors which originate from and involve the substance of the cord itself, and *extramedullary* tumors which occur in the meninges and the surrounding tissues. The intramedullary tumors are most frequently ependymomas and less often multiform glioblastomas and medulloblastomas. The extramedullary tumors include meningiomas, angiomas, lipomas, glands of Hodgkin's disease, tuberculomas, syphilomas, cysts of the spinal cord, and metastatic tumors An ependymoma of the filum terminale forms a giant-sized mass which compresses the roots of the cauda equina. Multiple neurofibromata of von Recklinghausen's disease may form in the cauda equina. The extramedullary tumor is more frequent, and the vast majority are benign and accessible to surgery. Intramedullary tumors occur most often in the cervical and the lumbar enlargements. A glioma of the cervical enlargement

[7] Boshes, B., A Review of Medicine, ed 6, Chicago, Northwestern University Medical School, 1951.

may become cystic and produce a syringomyelic cavity.

Symptoms

Symptoms are caused by direct pressure upon the cord and the nerve roots, pressure against the opposite side of the cord which is pushed against the bony wall, and by changes in blood vessels with altered blood and spinal fluid flow.

Pain[8] is the most frequent symptom and usually is felt in the region supplied by the involved posterior root. Thus a neuralgic pain may extend down a limb or be situated about the chest or the abdomen as a girdle pain. The usual pain of an intraspinal lesion may precede any other symptom by months or years and may be constant or intermittent. *It characteristically is most pronounced at rest and is reduced in intensity by exercise.* It usually persists in a well-localized area because of definite nerve root involvement. Commonly, it is lancinating and aggravated by coughing, sneezing, lifting, and straining during bowel movement It invariably awakens the patient 4 to 6 hours after he has retired and often becomes so severe as to compel the patient to walk the floor or to sleep in a sitting position

Pain may be confined to the back where it is generally situated about the site of the lesion. However, involvement of the cauda equina anywhere in its course from the upper lumbar canal often causes low back pain. Referred pain from the cervical area involves the upper extremity. Within the thoracic spinal canal, a lesion at a definite vertebral level will involve the dermatome several segments below Thus, a neurofibroma at the level of the 5th thoracic vertebra often causes pain in the subcostal area of the abdomen, where it may be mistaken for gallbladder disease. Cauda equina tumors cause sciatica as well as low back pain, a ruptured disk often being diagnosed.

Involvement of a motor nerve may cause painful spasms of the muscle which it innervates

Motor Signs. Eventually, motor signs appear in the form of muscle weakness at the level supplied by the involved segment of the cord and usually on the same side Involvement of the pyramidal tracts causes a spastic paralysis with hyperactive deep reflexes, absent superficial reflexes, and presence of pathologic tendon reflexes below the level of the lesion. As spinal cord compression continues, the opposite side is implicated likewise.

When the cauda equina is involved, the paralysis is flaccid, and both knee and ankle reflexes are lost.

Sensory changes are varied. Compression of the posterior columns causes a loss of deep sensation, position and vibration sense. When the lateral columns are compressed, the spinothalamic tracts are affected, and pain and temperature sensation is lost on the opposite side below the level of the lesion.

Brown-Sequard Syndrome. When pressure of the tumor compromises the function of one half of the spinal cord, a syndrome results which corresponds to hemisection of the cord. Pain and temperature sense is lost on the opposite half of the body, and position, vibration, deep pain and light touch, as well as motor function, are lost on the ipsolateral side.

Hyperesthesia and Hyperalgesia. Because of irritation of the posterior roots, often these conditions are found at the level of the lesion

Sphincter weakness with incontinence of urine and feces often develops with severe compression Impotence is commonly associated.

Trophic ulcers and bedsores may occur.

Perspiration is generally reduced or lacking below the level of the lesion.

Special Localizing Signs:

Cervical Lesions—occipital pain, neck rigidity, pain in the upper extremities, paralysis of the diaphragm when C3 to C6 are involved.

First Dorsal Lesions—Horner's syndrome: enophthalmos, anhidrosis, and ptosis of the upper lid. (The opposite effect is secured when the lesion is irritative.)

Dorsal Lesions—paraplegia, most often in flexion, pain radiating to the chest and the abdomen anteriorly.

Lumbar Lesions—shooting pains down the legs, low back pain, loss of knee or ankle reflex, spastic paralysis below level of lesion.

Cauda Equina Lesions—sciatic pain, low back pain, loss of both ankle and knee reflexes, flaccid paralysis.

Conus Lesion—severe sphincteric weakness and impotence, sad" anesthesia due to in-

[8] Craig, W McK. Need for consideration of intraspinal tumors as a cause of pain and disability, J A.M.A. 163:436, 1957.

volvement of the lowest sacral and coccygeal nerves.

Diagnosis

Spinal Fluid Examination. Changes in intraspinal pressure and chemical composition of the fluid are essential to diagnosis.

1. **Manometric Test.** The patient is placed in the lateral recumbent position, a lumbar puncture is performed, and a manometer is attached to the needle. The following observations are made:

A. *Initial Pressure*—Normally 80 to 150 mm.

B. *Oscillations*- -These are of two types: a slow rise and fall of the fluid level corresponds to respirations; a more rapid, smaller rise and fall occurs coincident with the pulse.

C. *Jugular Compression*—An assistant compresses both jugular veins momentarily. Normally, this causes an immediate rise of the column of spinal fluid which varies from 220 to 400 mm., followed by an immediate fall to the initial level. A space-consuming intraspinal lesion interferes with the free flow of spinal fluid as well as the free flow of blood through the veins of the spinal cord and roots. Increase of intracranial tension by jugular compression will not cause the normal rise in spinal fluid pressure when a mass is interposed between the cranium and the lumbar sac. Jugular compression will not affect the normal height of rise and, upon release of compression, the fluid level falls slowly.

D. *Abdominal Straining*—The patient strains momentarily as though at stool. This causes an immediate high rise and fall, regardless of the presence of a tumor.

E *Removal of Fluid for Examination*

2. **Spinal Fluid Changes.** There is a marked increase of albumin content just below the level of the lesion and sometimes just above the level. The total protein is much greater than that found with disk rupture. The diagnostic *Froin's syndrome* may appear below the level of the lesion: xanthochromia, spontaneous coagulation and no cells.

Roentgenologic Findings. Localized erosion and demineralization of adjacent bony structures may be found. Attention is directed especially to the pedicles which may be flattened in contour, the width of the spinal canal being correspondingly wider at this level. Large expanding lesions such as an ependymoma of the cauda equina are likely to effect bone changes. Destruction of an adjacent vertebra may indicate a malignant metastasis. Pantopaque injecion helps to localize the lesion. Characteristic shadows produced by certain tumors have been described but are not reliable.

Importance of a General Examination. Carcinomas of thyroid, breast, prostate and adrenal gland are prone to metastasize to the spinal column. These sites should be investigated thoroughly before exploration of the spine is attempted.

Treatment

Surgery aims at relief of compression of the spinal cord. The mortality rate is less than 4 per cent. Most tumors are extramedullary and, after their removal, symptoms subside to a remarkable degree. Intramedullary tumors are surgically inaccessible. Even when the lesion cannot be removed, laminectomy reduces pain and prolongs life by relief of pressure. It often prevents bladder paralysis and ascending urinary infection.

FILUM TERMINALE SYNDROME
(Cord Traction Syndrome)

Progressive spastic paralysis can occur in Arnold-Chiari syndrome, scoliosis and other spinal malformations. The mechanism appears to be an abnormally short filum terminale which pulls the cord distally as the vertebral column grows in length. In Arnold-Chiari syndrome, the hindbrain is pulled into the narrow foramen magnum. In scoliosis, the cord is pulled over the angulation. Typically, symptoms appear during periods of rapid growth, from 13 to 19 years of age. Shortened cauda equina is a complication particularly of spina bifida and causes retention of the infantile position of the conus medullaris. Treatment is by sectioning the filum, which gradually improves the patient. In Arnold-Chiari syndrome, decompression of the foramen magnum produces the same result.[9, 10] This latter condition consists of herniation of

[9] Garceau, G. J · Filum terminale syndrome, J Bone & Joint Surg 35A·711, 1953.

[10] Lichtenstein, B. W.: Distant neuroanatomic complications of spina bifida (spinal dysraphism), hydrocephalus, Arnold-Chiari deformity, etc : pathogenesis and pathology, Arch. Neurol & Psychiat. 47:195, 1942.

the hindbrain and the cerebellum through a narrowed foramen magnum. It occurs during the growth period when the spinal cord is prevented from ascending by a tight filum terminale. The constriction of the hindbrain interferes with the exit of spinal fluid from the ventricles, and hydrocephalus results in infants, and signs of increased intracranial pressure and spastic paralysis in older children. In addition, the discrepancy in growth between the spinal cord and the spinal column results in reversal in the course of the cervical spinal nerve roots which become angulated over their point of exit at the intervertebral foramina.

When progressively increasing weakness in the lower extremities, deformities of the feet, sphincter weakness, root pains in the upper extremities and headaches develop in an actively growing youngster, a tight filum terminale and the Arnold-Chiari deformity should be suspected. The fibrous tissue formation about a spina bifida occulta is often the aggravating factor (see Spina Bifida).

SPINAL MUSCULAR ATROPHIES OF UNKNOWN ETIOLOGY

The importance of these diseases to the orthopaedic surgeon lies in differentiating them from poliomyelitis. Certain of these conditions, such as peroneal atrophy and Friedreich's ataxia, are benefited by surgical treatment.

Progressive Spinal Muscular Atrophy

This disease is characterized by progressively increasing weakness and atrophy which usually begin in the small hand muscles and extend to the arms and the lower extremities The cause is unknown

Pathology. The cervical spinal cord is affected most severely, but eventually all portions become involved. The anterior horn cells degenerate, disappear and are replaced by glial tissue. Occasionally, the medulla may be involved.

Clinical Picture. Symptoms develop insidiously in a middle-aged individual, usually a male. *Weakness and awkwardness in use of the hands is often the first complaint* Pain is absent The small muscles of the hand atrophy, producing a characteristic clawhand. Gradually, the weakness and the atrophy increase in degree and spread to the arms and the shoulders and after a long period of time involve the legs. The process may involve one hand at first but eventually becomes symmetric. It rarely starts in the lower extremities but invariably spreads to the upper extremities.

Examination reveals atrophy, muscle fasciculations, absent deep reflexes, and a reaction of degeneration. Sensation is intact.

Prognosis. The course is slow and protracted, often as long as 25 years. Rarely is death caused by bulbar symptoms.

Familial Progressive Spinal Muscular Atrophy (Werdnig-Hoffmann Disease)

This is a rare form of progressive degeneration of the anterior horn cells. It is characterized by a familial incidence, onset of flaccid paralysis in early infancy, and progression to a fatal termination in about 5 years. Symptoms typically start in the trunk and spread peripherally along the extremities.

Amyotonia Congenita (Oppenheim's Disease)

In this condition the anterior horn cells, particularly those in the lower half of the spinal cord, have failed to develop. As a result the lower extremities of the newborn infant display a flaccid weakness or paralysis with absent deep reflexes The condition tends to improve a little with passage of time.

Amyotrophic Lateral Sclerosis

This is characterized by rapid progression of degeneration of the anterior horn cells and pyramidal tracts involving both the spinal cord and the brain stem. It appears most often in middle-aged males. Symptoms include weakness, atrophy and flaccid paralysis in the upper extremities, spastic paralysis in the lower extremities, and bulbar paralysis, which at first is manifested as difficulty in swallowing and talking Atrophy involves all muscles. The gait is typically spastic. Sensory disturbances are absent. The disease is rapidly progressive, death occurring in 2 to 5 years in most instances.



FIG. 138. Charcot-Marie-Tooth peroneal muscular atrophy. Note the "stork legs" and pes cavus. (Case of Dr. Alexander T. Ross)

PERONEAL MUSCULAR ATROPHY (Charcot-Marie-Tooth Disease; Hereditary Muscular Atrophy)

This hereditary disease is characterized by slowly progressive symmetric muscular atrophy and weakness of the legs and the feet with eventual involvement of the forearms and the hands. Involvement of the peroneal muscles is typical.

Etiology. The cause is unknown. Males are affected more often than females. The disease is transmitted as a dominant, recessive, or sex-linked characteristic.

Pathology. A degenerative process without an inflammatory reaction affects the anterior horn cells and the peripheral nerves and, to some extent, the posterior columns

Clinical Picture. The onset of symptoms tends to appear at about the same age in childhood in the afflicted members of the family. Initially, the gait is clumsy, and peroneal weakness is demonstrable in frequent ankle inversion strains. Pains and paresthesias are common in the legs. Very slowly over many years there develops the typical picture of atrophy of the leg muscles and the intrinsic muscles of the feet. At first a foot-drop and inversion of the ankle develop into a fixed equinocavovarus deformity with clawed toes. The gait is steppage. The peroneals usually show the greatest amount of weakness. A comparable muscle atrophy and deformity later develop in the upper extremity, causing thinning of the forearms and clawed hands. The deep reflexes are lost. Loss of tactile, temperature and proprioceptive sensations is variable in degree. The face, the trunk, the pelvic, and the shoulder girdles are not affected.

Prognosis. The course is slow and protracted but may become stationary at any time The patient may live out the normal life's expectancy.

Treatment. Prostigmine bromide (15 mg t.i.d.) is often helpful in lessening weakness

and ataxia. A spring drop-foot brace and dynamic splinting of the hand prevent contractures. When deformity is extreme, surgical procedures are necessary. The foot may be corrected by a panastragalar arthrodesis plus tarsal wedge resection to overcome the cavus. Heel-cord lengthening alone is of no value because the deformity progresses. Surgical procedures are also available for overcoming intrinsic muscle paralysis and contracture (see section on The Hand).

Friedreich's Ataxia (Hereditary Spinal Ataxia)

This hereditary and familial disease is characterized by progressive degeneration of the corticospinal, the spinocerebellar and the posterior columns of the spinal cord.

Etiology. The cause is unknown. Transmission occurs through affected and unaffected individuals.

Pathology. A demyelinizing process occurs in the pyramidal tracts, the spinocerebellar tracts and especially the posterior columns. When destruction is severe and extensive, the anterior horn cells may also be involved.

Clinical Picture. The average age of onset is 10. Initially, the complaints include awkwardness of gait, stumbling, frequent falling, frequent twisting of the ankles, foot fatigue and difficulty in obtaining comfortable shoes The shoes wear out rapidly and become misshapen as foot deformity gradually develops. Typically, a symmetric clawfoot forms with marked elevation of the longitudinal arch, prominent metatarsal heads, widening of the fore part of the foot, and hyperextension and clawing of the lesser toes. An equinus is only apparent and is due to dropping of the fore part of the foot at the midtarsal area Muscle weakness becomes apparent in the peroneals and the anterior tibials. This clinical picture is quite common and may not progress beyond this point, thereby constituting an abortive form of the disease.

In typical Friedreich's ataxia, symptoms and findings slowly progress over the years. The gait becomes ataxic, and Romberg's sign is positive. Cerebellar signs include failure to perform the finger to nose test and adiodokokinesis. Deep pain, position and vibration sense are impaired, attesting to a posterior column lesion. The deep tendon reflexes are lost early. Later, as the pyramidal tracts are involved, the reflexes may become hyperactive, and the Babinski sign is positive. Speech becomes halting and explosive. Muscle weakness and atrophy affect particularly the intrinsic muscles of the feet and the leg, especially the peroneals and the anterior tibials. An extreme equinocavovarus deformity may develop and in itself, regardless of the ataxia, is very disabling. Eventually, usually at about the age of 30, the patient becomes bedridden. A kyphoscoliosis is occasionally an additional deformity.

Treatment.[11] Surgical intervention is indicated both for the mild abortive form and the severe type of the disease. In the latter, provision of a stable, corrected foot will greatly aid ambulation and may add many useful years to these individuals.

The basic procedure consists of a triple arthrodesis with adequate wedging resection to overcome deformity. Because the equinus is limited to the fore part of the foot, Achilles tendon lengthening is contraindicated. Instead, the midtarsal resection and subcutaneous fasciotomy, if necessary, will suffice. When anterior tibial function is inadequate, the extensor hallucis longus is transferred to the neck of the first metatarsal, and the interphalangeal joint of the large toe is fused. Bone surgery should be postponed until bone maturity is adequate.

Syringomyelia

Syringomyelia is a slowly progressive disease of the spinal cord and the medulla oblongata caused by cavitation and gliosis and characterized clinically by dissociated sensory loss and muscular atrophy.

Etiology. The cause of true syringomyelia is unknown. Pseudosyringomyelia is cavitation which is localized and secondary to vascular lesions, intramedullary tumors, or trauma to the cord.

Pathology.[12] The essential lesion is the slowly progressive destruction of the central portion of the cervical enlargement of the spinal cord from where it extends downward to involve

[11] Makin, M · The surgical management of Friedreich's ataxia, J. Bone & Joint Surg. 35A·425, 1953

[12] Lichtenstein, B W.. A Textbook of Neuropathology, p 224, Philadelphia, Saunders, 1949.

the thoracic and the lumbar segments and upward into the medulla oblongata. The central lesion is a cavitation which extends into the posterior and the anterior gray columns and then compresses and sometimes destroys the posterior and the lateral white columns. Microscopically, abnormal masses of ependymal cells lie in close relationship to thin-walled blood vessels, suggesting a choroid plexus which secretes cerebrospinal fluid. It is the accumulated fluid which forms the cavity and extends along lines of least resistance. When the choroid plexuslike structure is destroyed, fluid no longer forms, and progress of the degenerative process is arrested. Similarly, surgical drainage of the syrinx with provision of a permanent external communication, or spontaneous perforation into the central canal stops progress of the disease.

Clinical Picture. The onset is insidious. Weakness and atrophy of the intrinsic muscles of the hands are often the initial symptoms and findings. Pain—sharp shooting or burning—throughout the upper extremities is common. The loss of painful sensation is first disclosed by unnoticed burns and injuries. Examination reveals loss of pain and temperature sense, usually extending in a shawllike distribution over the arms and the shoulders. Tactile sensation is undisturbed. Reduced sensation to touch, when present, is associated with loss of proprioception and signifies extension of the lesion into the posterior white columns. When spasticity, hyperactive reflexes and pathologic reflexes are present in the lower extremities, the lesion has compressed the pyramidal tract laterally. Progression of the lesion may halt at any time. Involvement of the medulla oblongata (syringobulbia) is indicated by aphonia, dysphagia, etc. Charcot's joint develops in approximately 20 per cent of cases in the upper extremity. Scoliosis is a frequent deformity

Prognosis. Although slowly progressive, the disease may become stationary at any time. Bulbar involvement is of serious import

Treatment. Drainage of the cavity is indicated. A platybasia with an Arnold-Chiari deformity is a not uncommon cause of pseudosyringomelia This should be investigated and decompressed if necessary. A Charcot joint at the shoulder or the elbow may require stabilization.

DIFFERENTIAL DIAGNOSIS OF THE MUSCLE ATROPHIES

1. **Progressive Spinal Muscular Atrophy.** Upper extremity, especially the hand, is chiefly affected, is painless, sensation is intact, onset in middle age, very slow progression.

2. **Peroneal Atrophy.** Condition familial, several members of family afflicted, lower extremities earliest and chiefly involved, shoulder and pelvic girdles escape involvement, pain often associated, peroneal paralysis is characteristic, onset is in childhood. Rarely fatal.

3. **Familial Progressive Spinal Muscular Atrophy.** Condition familial but has onset in early infancy, starting in the trunk and progressing rapidly to involve the extremities. Fatal termination in a few years.

4. **Amyotonia Congenita.** Flaccid paralysis of lower extremities is already present at birth, tendency toward some improvement.

5. **Amyotrophic Lateral Sclerosis.** Onset in middle age of flaccid paralysis in upper and spastic paralysis in lower extremities, rapidly progressive, involving medulla. Fatal in a few years.

6. **Syringomyelia.** Atrophy is associated with loss of pain and temperature sense and preservation of tactile sense in the upper extremities.

7. **Poliomyelitis.** Preceded by a febrile disturbance, paralysis often transient in some areas, atrophy is asymmetric, spotty and nonprogressive.

8. **Cervical Cord Tumor.** Root pain and atrophy and may be unilateral, progressively worse, sensory impairment affecting all forms, headache common, spinal fluid studies may show a subarachnoid block and increase of total protein.

9. **Friedreich's Ataxia.** The muscle atrophy affects the leg and the foot, causing a typical severe equinocavovarus deformity, onset in childhood, ataxia, loss of proprioception, pyramidal tract signs, hereditary and familial.

PERIPHERAL NERVES

Function of a Peripheral Nerve

Peripheral nerve fibers conduct sensory, motor and trophic impulses. Sensation includes coarse and light touch, pain, temperature,

stereognosis and deep tissue sense (pain, position, vibration). Motor fibers innervate muscles. Trophic fibers are supplied to all tissues including skin, tendons, joints and muscles.

Basic Nerve Unit

The motor fibers originate from neurons in the anterior horn of the spinal cord. Sympathetic neurons in the lateral columns of the gray matter give rise to vasomotor and trophic fibers. All fibers combine and emerge as myelinated fibers from the anterolateral aspect of the cord as a common white ramus. A ganglion located outside the dorsolateral aspect of the cord is connected with the latter by a dorsal gray nerve root. It contains the neurons for sensory perception.

Pathology Following Severance of a Peripheral Nerve

All functions distal to the point of severance are interrupted. At the end of the proximal nerve segment the axons multiply and attempt to grow distally. However, a connective tissue bulblike growth envelops the end of the nerve and obstructs the path of these fibrils which become arranged in disorderly fashion. The connective tissue and fibril growth is called a neuroma. The distal nerve segment swells to twice its original size and undergoes wallerian degeneration. This process is complete in about 1 month. Its proximal end displays only a small enlargement, consisting only of fibrous tissue. Occasionally, the connective tissue growths at the end of each segment may unite, and some of the fibrils may successfully penetrate the mass and grow distally. Partial function may thereby be restored.

Symptoms and Findings Following Nerve Injury

The following functions are lost·

Stereognosis, the most specialized perception of shape and texture. The specialized touch corpuscles (Pacini's, Ruffini's, Meissner's) located in the hand, most particularly in the median nerve distribution, are linked with the stereognostic center on the opposite side of the brain.

Superficial sensation to touch, pain and temperature includes epicritic sensation (light sensation) by which two points of a compass are distinguished, and coarse sensation such as pain.

Deep sensation to muscle and joint movements, position, deep pressure and vibration travels mainly in motor nerves. Bunnell describes an excellent method of mapping anesthetic areas by an electric skin resistance machine consisting of a battery, a galvanometer and 2 electrodes placed near one another. Anesthetic skin is electroresistant because the sweat glands are dry. Placed on normal skin, this apparatus is a detector of malingerers as the galvanometer will show normal conductivity. An individual who claims to have pain will show excellent conductivity because pain stimulates the sweat glands. No reaction is obtained on anesthetic skin.

Loss of motor supply to a muscle results in progressive atrophy and fibrous degeneration of that muscle. A muscle is partially paralyzed when nerve severance is incomplete and is revealed by a limited amplitude of motion and decreased force against resistance.

Deep reflexes are diminished and lost.

Electric stimulation of the nerve no longer causes the muscle to contract. However, the muscle may be well stimulated directly by faradic current. This response gradually diminishes until after 2 weeks no response to faradism is obtainable. Nevertheless, the muscle continues to respond to galvanic current by a slow vermicular contraction, greater in amplitude, and followed by slow relaxation. This chain of events, namely, early loss of response to faradism and increased continued response to galvanism, is known as the *reaction of degeneration* and is characteristic of peripheral nerve interruption. After the muscle has undergone complete fibrous degeneration no further electric reaction is obtainable. Each muscle responds best electrically at the point where the nerve enters the muscle. Normally, it contracts strongly to faradic current and gives a quick twitch to galvanic.

Trophic influence is lost. All tissues in the supplied area undergo atrophy. The skin is thin and glossy, red or cyanotic. Hair and nails are brittle. Bone is osteoporotic. Joint cartilage is thinned, and ligaments are contracted and inflexible, resulting in decrease of motion and fibrous ankylosis of the joint. Healing of wounds is slow. This picture should be differentiated from the condition of reflex

sympathetic dystrophy characterized by a generally painful, swollen, cold, cyanotic part and typical mottled osteoporosis.

Determining the Site of Nerve Injury

The particular nerve involved is revealed by the muscles paralyzed and the area of anesthesia. The point of interruption is located by the history of accident, location of the nerve and by Tinel's sign. The last is performed by percussing or tapping over the severed nerve end, causing tingling in the area of distribution of the nerve. In the course of regeneration the sign can be elicited further distally, indicating the level to which the new axons have grown.

Regeneration of Nerves

Severed nerves will bridge a gap of 1 cm. or a little more. At operation a nerve stripped of its surrounding tissues loses its blood supply and function temporarily. Scar tissue will strangulate a nerve and obstruct peripheral growth. Repair of a peripheral nerve demands accurate approximation in exact axial rotation; otherwise, sensory fibers may grow down motor pathways, and vice versa, and so are wasted. Regeneration occurs at a rate of 1 or 2 mm. a day. Motor recovery can occur up to 2 or 3 years, and sensory return from 3 to 5 years after nerve severance In the arm, the radial nerve regenerates better than the median; and the median better than the ulnar. Sensation recovers before motor function Protopathic precedes the epicritic sense. Deep sensibility returns with the epicritic and, finally, stereognosis. The proximal portions of the anesthetic area disappear first. Tinel's sign can be elicited by tapping anywhere over the newly formed nonmyelinated axons. The sign disappears in 1 or 2 years as the axons become myelinated. The quality of sensation early after return is not normal. Paresthesia is felt in response to stimuli. Reactivation of muscles occurs later, the most proximal ones returning first The early flicker of motion increases until a large portion of the muscle contracts, although it moves the part through a limited amplitude and is easily fatigued. Strength and co-ordination in movement are acquired eventually Only after voluntary motion reappears does the reaction to electric stimuli return. Galvanic reaction returns before faradic as a rapid response of the muscle, not the slow, vermicular contraction. Return of faradic reaction may never occur or is weak. Trophic changes progress even after nerve severance; these start to regress when sensation begins to appear.

The Degree of Nerve Injury

Neurapraxia is a physiologic injury to a nerve. No anatomic damage is present. The paralysis is transient, sensory loss is slight, no reaction of degeneration is obtainable, and recovery is complete within a few hours to days.

Neurotmesis is complete physiologic and anatomic interruption of the nerve fibers and their sheaths. Recovery generally is obtainable by surgical approximation.

Axonotmesis is interruption of nerve fibers within their sheath. The Schwann tubes remain in continuity so that spontaneous cure eventuates. It is necessary to distinguish between neurotmesis and axonotmesis to determine whether to intervene surgically.

Traction nerve injuries are often severe and are essentially a neurotmesis. Although anatomic continuity may appear to be preserved, extensive intraneural scar formation occurs. Direct compression injuries at a fracture site usually have a good prognosis for spontaneous cure. This is especially so in the case of the radial nerve. Involvement of the axillary nerve has an unfavorable prognosis. It is necessary to preserve muscle and joint function by galvanic stimulation, passive motion and daily massage until the nerve regenerates. Inasmuch as axons grow down 1 mm. per day, one can estimate the time required to reach the most proximally involved muscle; e g., the brachioradialis in case of the radial nerve. If reinnervation does not occur at the expected time, surgical exploration of the nerve should be undertaken. An electromyographic study of motor unit action potentials is found in the most proximal muscle some weeks before a flicker of voluntary power is ascertained clinically.

Multiple Neuritis[13]
(Polyneuritis)

The term "neuritis" is applied to a painful,

[13] Boshes, B.: A Review of Medicine, ed. 6, Chicago, Northwestern University Medical School, 1951.

TABLE 5. PRINCIPAL CAUSES OF NEURITIS

GENERALIZED POLYNEURITIS			
A. Virus	*B. Bacteriotoxic*	*C. Deficiency or Metabolism*	*D. Chemical*
Measles	Focal infections	Pellagra	Mercury
Smallpox	"Rheumatism"	Pernicious anemia	Lead
Chickenpox	Erysipelas	Sprue	Silver
Parotitis	Scarlet fever	Beriberi	Arsenic
Herpes	Rheumatic fever	"Alcoholic neuritis"	Phosphorus
"Acute febrile"	Chorea	"Korsakow's psychosis"	Methyl alcohol
"Acute infective"	Septicemia	Pernicious vomiting	Ethyl alcohol
"Landry's"	Puerperal fever	Hunger edema	Ethyl iodide
Poliomyelitis	Gonorrhea	Pregnancy	Trichlorethylene
Encephalomyelitis	Meningitis	Chronic colitis	Carbon tetrachloride
Epidemic (lethargic) encephalitis	Diphtheria	Cancer with cachexia	Trinitrotoluene
	Typhoid fever	Tuberculosis with cachexia	Dinitrobenzene
Erythro-edema	Paratyphoid fever	Senility with cachexia	Triorthocresyl phosphate
Acute rabic myelitis	Typhus fever	Diabetes	Aniline
	Influenza	Myxedema	Sulfonethylmethane, barbital, etc.
	Pneumonia	Hematoporphyrinuria	
	Malaria	"Recurrent polyneuritis"	Chloral, Chlorbutanol
	Relapsing fever	"Chronic progressive polyneuritis"	Carbon monoxide
	Serum sickness		Carbon bisulphide
	Acute enteric fever	Chronic bacillary dysentery	

LOCALIZED NEURITIS	
A. Mechanical	*B. Infectious*
Pressure	Diphtheria
tumor	Tetanus
edema	Streptococci
arthritis	Leprosy
fibrosis	
Trauma	
Saturday night paralysis	
Volkmann contracture	
Meralgia paresthetica	

degenerative, often inflammatory process in a neuron and its fiber. When any portion of a neuron, whether axon or cell body, is involved, the remainder of the cell invariably undergoes changes. Therefore, the painful degenerative process, regardless of its initial situation, causes functional loss of the entire unit.

Neuritis is very common and is caused by a great many conditions. In Table 5, the principal causes are divided into those causing mononeuritis and others causing polyneuritis. Neuritis of a single nerve most often stems from local causes which are described under their respective sections. The following discussion applies mainly to multiple neuritis

Pathology. Mild damage to a peripheral nerve causes swelling of the axon sheath and is reversible. With more intense trauma sheath, axon and myelin disintegrate. Regeneration takes place at the rate of 1 mm. per day.

Clinical Picture. Symptoms pertain to both motor and sensory divisions of the peripheral nerve.

1. SENSORY SYMPTOMS. *Pain* is common and often severe. It varies in intensity and character and may be sharp, burning or boring It always follows the course or distribution of the nerve and disappears when the nerve is severed at its proximal portion. *Paresthesias* often precede and follow the pain. The nerve is tender along its entire course.

2. SENSORY FINDINGS. All forms of sensation, including pain, touch, temperature, position and vibration, are reduced or entirely lost.

3. Motor Loss. Impairment varies from paresis to paralysis. Muscles are flaccid and lack the resiliency of muscles with tone. Atrophy develops. The deep reflexes are reduced or absent.

4. Electric Reactions. Reaction of degeneration develops.

5. Cramps and Muscle Spasm. Occasionally occur at the onset because of an irritative lesion.

6. Trophic and Vasomotor Changes. Sympathetic and trophic fibers run through the peripheral nerve. The skin becomes thin, glossy, cyanotic and cold. Hyperhydrosis or hypohidrosis, hypertrichosis and nail changes may occur.

7. Muscular and Joint Contractures. May develop as a result of unbalanced muscle pull.

Acute Infectious (Febrile) Polyneuritis. This is composed of a large group of so-called "virus diseases," including measles, smallpox, herpes zoster, parotitis, etc. Epidemic (lethargic) encephalitis must be included because the disease begins so often with muscle tenderness, flaccid paralysis and paresthesias. These virus diseases not only affect the peripheral nerve but also the anterior horns, the spinal ganglia and white and gray matter of the cortex.

Clinical Picture. The onset is acute or subacute with fever, chills, malaise and anorexia. Several weeks later the signs of polyneuritis appear. Pains appear in the back and the extremities. The arms and the legs feel weak and shaky, and a flaccid paralysis gradually develops, followed by numbness and tingling. Usually, there is bilateral facial weakness, together with mild sphincteric disturbances. Nerve and muscle tenderness is prominent All forms of sensation are diminished

Bacteriotoxic Polyneuritis. Many bacterial diseases are often associated with a transient peripheral neuritis. The mechanism of causation may be bacterial, toxic and lack of vitamins. The following are especially important.

1. Septicemic Polyneuritis. Symptoms are similar to those caused by alcohol. In addition to pain, tenderness and hyperesthesia of the skin, paresthesias are frequent, and trophic changes of the fingers occur. The cranial nerves may be involved.

2. Diphtheritic Polyneuritis. Cranial nerve involvement occurs within a few weeks after the sore throat. Bulbar symptoms include aphonia, nasal regurgitation of liquids, dysphagia, and abductor paralysis of the vocal cords. Facial and ocular paralysis, limb paralysis, particularly of the extensor muscles, and respiratory and cardiac paralysis may develop. The paralysis usually develops first in the vicinity of the diphtheritic membrane—hence the frequency of cranial nerve involvement. Spread of infection is apparently via perineural lymphatics. The palatal, pharyngeal and laryngeal paralysis clear up in a few weeks; ocular palsies last longer; while those of the extremities last a long time.

3. Enteric and Paratyphoid Polyneuritis. Neuritis is asymmetric and often starts with severe pains about the shoulders, followed by atrophy of the trapezius or the serratus magnus. Dietary vitamin deficiency is the probable cause.

4. Serum Disease Neuritis. Whether the serum or the treated disease is the cause is debatable.

5. Puerperal Polyneuritis. This occurs several weeks before term. The causative factor is prolonged avitaminosis associated with hyperemesis gravidarum.

6. Rheumatic Polyneuritis. This designation is given to a mild type of polyneuritis associated with rheumatoid arthritis. It generally disappears with improvement of the arthritis, leaving atrophic stiff muscles. Intrinsic muscle contracture of the hand, a frequent residual of rheumatoid arthritis, may very well be the result of neuritis.

Deficiency or Metabolism Polyneuritis. Some unknown factor causes inflammatory changes in the peripheral nerves and degenerative changes in the spinal cord. The mechanisms at work include lack of digestive juices in the stomach, lack of vitamin B, failure of absorption from the gastro-intestinal tract, and inadequate diet.

1. Beriberi. This disease, formerly seen in India and Japan, is due to thiamine deficiency. The onset is acute with cardiac dilatation and tachycardia but no fever. Edema is prominent about the body but chiefly affects the lower extremities. Nausea, vomiting and depression are followed by polyneuritic pains, arreflexia, sensory disturbances and muscle atrophies. The course lasts from a few weeks to several months, and death occurs from cardiac failure or intercurrent disease. Edema may be absent.

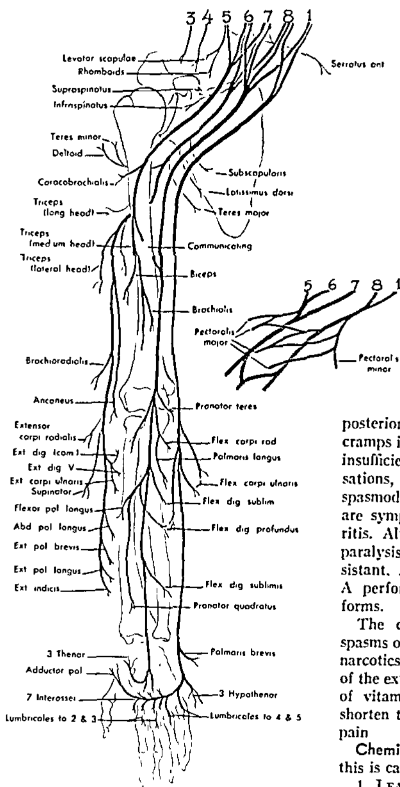


Fig. 139. Nerve supply to muscles of the upper extremity.

Subclinical vitamin B_1 deficiency develops with dietary inadequacy, anorexia, failure of assimilation or increased requirement in thyrotoxicosis, pregnancy and lactation

2. Diabetic Polyneuritis. This occurs in individuals above 40 as an interstitial inflammatory neuritis and degeneration in the posterior roots with secondary changes in the posterior columns. Dull aching pains and cramps in the legs are often due to circulatory insufficiency. On the other hand, burning sensations, hyperesthesias of the skin, sharp, spasmodic, lightninglike pains, and numbness are symptoms characteristic of diabetic neuritis. Although the deep reflexes disappear, paralysis is rare. Pain is distressing and resistant. Ataxia may resemble tabes dorsalis. A perforating ulcer of the foot sometimes forms.

The course is generally one of painful spasms occurring over several weeks, requiring narcotics and then subsiding. Immobilization of the extremity in plaster, plus administration of vitamin B_{12} in large amounts seems to shorten the course and lessen the intensity of pain

Chemical Multiple Neuritis. Most often, this is caused by lead, arsenic and alcohol.

1 Lead Neuritis. Lead poisoning differs from other forms of polyneuritis in causing involvement of the central nervous system, especially the anterior horn cells. Painters, plumbers, plasterers, and typesetters are predisposed. Cosmetics, hair dyes and many other substances contain lead

The degenerative neuritis is limited to motor nerves, especially those to the extensors of the hand and the forearm. However, the fore-

arm flexors and the deltoid finally are involved. Usually, the brachioradialis and sometimes the extensor and the abductor of the thumb are spared. When the lower extremity is involved, the peroneals are more likely to be affected than the anterior tibial. The course is chronic, and the outlook is good when the offending agent is removed.

2. Arsenical Polyneuritis. The source of arsenic includes medications, wines and insecticides. Symptoms resemble those of alcoholic polyneuritis. Pain in the limbs and numbness of the hands and the feet are followed by wrist-drop and foot-drop. The muscles atrophy rapidly. Cutaneous anesthesia, pains and hyperesthesias are more severe than in alcoholic polyneuritis, and mental symptoms are rare.

Associated symptoms of arsenical poisoning include abdominal pain, vomiting and diarrhea, skin pigmentation, keratoses and herpes zoster.

The diagnosis is established by recovering arsenic from the urine and the hair.

Recovery requires from 2 to 3 years.

3. Mercurial Polyneuritis. This resembles other forms, but renal damage and gingivitis are additional findings.

4. Alcoholic Polyneuritis. Alcohol acts not only directly on neural tissue by coagulating the albumin but also indirectly by creating a dietary deficiency and avitaminosis. The latter mechanism, which also operates in pellagra, causes not only polyneuritis but also extensive nervous tissue damage with degeneration of the posterior columns of the spinal cord and lipochrome deposits.

The onset displays fever, muscle tenderness, pain in the limbs, and cutaneous hyperesthesia. Delirium and insomnia are frequent. Tingling, numbness and loss of pain sensation develop. The foot dorsiflexors weaken and foot-drop is apparent. Tenderness of the nerve trunks is revealed, especially over the peroneal and the ulnar nerves. Wrist-drop and generalized muscle weakness may occur in severe cases. Trophic changes in the nails and the skin of the fingers are seen. Sweating of the hands and the feet and edema of the ankles are very common. Mental symptoms, the so-called "Korsakoff's psychosis," is sometimes associated and may persist after the neuritis has subsided.

The course is chronic, lasting a few months to 2 years. High dosage of vitamin B is beneficial.

Meralgia Paresthetica
(Lateral Femoral Cutaneous Neuropathy)

The lateral femoral cutaneous nerve arises from the posterior divisions of the 2nd and the 3rd lumbar nerves. It makes its appearance at the lateral border of the psoas and passes obliquely across the iliacus to the anterior superior iliac spine, where it proceeds beneath the inguinal ligament to enter the anterolateral aspect of the thigh

Any syndrome manifesting numbness, paresthesias and pain over the lateral and anterolateral aspect of the thigh suggests an inflammatory, degenerative lesion of the lateral femoral cutaneous nerve and is designated as *meralgia paresthetica*. The sensations experienced are described variously as burning, tingling, hyperesthesia, numbness, or severe pain. Often pain occurs after activity or direct pressure against the thigh and is relieved by rest. Reduction of tactile sensation is demonstrable over the lateral aspect of the thigh.

The cause is unknown. An osteoarthritic irritative lesion at the intervertebral foramina has been blamed.

Treatment. Symptoms may persist for many months but invariably subside. If pain is intolerable, removal of the cutaneous nerve may be tried. This can succeed only if pain is relieved by injecting a local anesthetic about the nerve at its point of emergence from the pelvis. Otherwise, the lesion must presumably be located proximal to this point.

INDIVIDUAL PERIPHERAL NERVES

Axillary Nerve

C5-

Anatomy. This nerve springs from the posterior cord of the brachial plexus and runs alongside the radial nerve behind the axillary artery, separating the latter from the subscapularis muscle on the floor. At the lower border of the muscle, it leaves the radial nerve and turns posteriorly, in company with the posterior humeral circumflex artery, through the quadrangular space to the posterior aspect of the humerus, where it divides into anterior and posterior branches. The posterior branch

gives off the nerves of supply to the teres minor and the posterior part of the deltoid before it curves around the posterior border of the deltoid to supply the skin over the lower half of the deltoid (upper lateral cutaneous nerve of the arm). The anterior branch proceeds laterally and anteriorly beneath the deltoid and in contact with the surgical neck of the humerus about 2 inches below the upper attachment of the deltoid, giving off numerous twigs to the muscle throughout its course The nerve of supply to the shoulder joint originates from the main nerve in the quadrangular space.

Clinical Picture. Injury to the axillary nerve occurs by a direct contusion or actual severance of nerve fibers in the course of surgical reflection of the deltoid from its attachment to the acromion, the clavicle and the scapular spine. Involvement of the main nerve in the axilla or at the surgical neck of the humerus produces complete paralysis of the deltoid with loss of abduction at the shoulder plus anesthesia of a small patch of skin over the lower half of the deltoid. The first 20° to 30° of abduction is initiated by the musculotendinous cuff, chiefly the supraspinatus component. The amount of this range of motion varies with the condition of the rotator cuff muscles. Occasionally, even with complete reaction of degeneration and atrophy of the deltoid, the supraspinatus may hypertrophy in compensation and restore abduction. When the anterior branch is interrupted, the muscle anterior to the point is paralyzed. Partial muscle paralysis is frequently compensated for by hypertrophy of the supraspinatus and the activity of the pectoralis major, especially with the arm above the horizontal plane.

Treatment. Prophylactic treatment consists chiefly of avoiding unnecessary extension of operative incisions and rough handling of the deltoid muscle. When the nerve has been contused, spontaneous regeneration may take place in 4 to 6 months, during which time the deltoid must be relaxed on an abduction frame and light massage and electric stimulation administered. Daily active exercises are done in an attempt to strengthen the cuff muscles. Operative repair of this nerve is exceedingly difficult—frequently impossible. Conservative treatment is advised unless severance of the main large nerve is evident.

Operative exposures of the shoulder should avoid muscle-splitting incisions. If unavoidable, the incision should be confined to the anterior third of the deltoid and not beyond 1½ inches distal to the acromioclavicular joint.[14]

ARTHRODESIS. Complete deltoid paralysis, when not compensated by other muscle action, requires arthrodesis of the shoulder. The trapezius and the serratus anterior will raise the arm effectively.

HARMON OPERATION.[15] When the anterior portion of the deltoid is paralyzed, the muscle may be seriously weakened, particularly in forward flexion, and the humeral head may even dislocate or subluxate anteriorly. This condition is remedied by transposing the posterior origin of the functioning muscle to a new anterior position.

TRAPEZIUS TENDON TRANSPLANT.[16] The trapezius muscle insertion is transferred by fascia lata strip extensions to the deltoid tubercle. The prerequisite is good power in all scapular muscles, including trapezius, serratus anterior, pectoralis major, rhomboidei and levator scapulae muscles. The main contraindication is subluxation of the shoulder joint

BICEPS AND TRICEPS TRANSFERENCE. The tendons of the short head of the biceps and the long head of the triceps are fixed to the anterior and the posterior rims of the acromion.

MEDIAN NERVE

Anatomy. The median nerve extends from the junction of the lateral head (from the anterior divisions of the 5th, the 6th and the 7th cervical nerves) and the medial head (from the 8th cervical and the 1st thoracic nerves). It enters the axilla laterally to the axillary artery and lies between the musculocutaneous nerve laterally and the ulnar nerve medially. The nerve descends in the arm with the brachial artery and other nerves in a groove just medial to and slightly behind the biceps muscle and gradually crosses over in

[14] Abbott, L C, *et al*: Surgical approaches to the shoulder joint, J. Bone & Joint Surg. 31A 235, 1949

[15] Harmon, P. H.: Surg, Gynec. & Obst. 84:117, 1947.

[16] Mayer, L: Dean Lewis Practice of Surgery, Hagerstown, Md, Prior, 1947

front of the artery (rarely, it crosses behind) until it lies medial to the artery before it reaches the elbow. The pulsations of the artery can be felt throughout its course in the arm and provides an excellent anatomic landmark for approaches to the nerve.

No branches are given off in the arm, except occasionally when the nerve to the flexors of the forearm has a high origin. At the elbow it lies deep to the bicipital aponeurosis (lacertus fibrosus) and the median cubital vein. It enters the forearm by passing between the larger humeral and the smaller ulnar head of the pronator teres, descending in the medial part of the forearm between the sublimis and the profundus muscles. Above the wrist it is radial to the sublimis and directly beneath the palmaris longus tendon. Then it passes beneath the transverse carpal ligament and after giving off the motor branch to the thenar muscles (opponens, abductor brevis, superficial head of the flexor brevis) it inclines volarward to supply by 6 terminal branches the thumb, the index, the middle and the ring fingers. In the hand it lies in a plane superficial to the tendons and deep to the superficial vessels, although its branches to the fingers are volar to the vessels.

The first branches arising from the nerve just above the elbow are those to the humeral head of the pronator teres. Then, below the elbow, branches supply the rest of the pronator teres, the flexor carpi radialis, the palmaris longus and the flexor digitorum sublimis. At the upper border of the pronator teres a large interosseous branch arises, also penetrates between the pronator heads, supplies the radial portion of the flexor profundus and the flexor pollicis longus, then descends on the interosseous membrane along with the anterior interosseous artery to end in the pronator quadratus.

In the hand, the median nerve gives off 5 palmar digital nerves. The first 3 supply both sides of the thumb and the radial half of the index finger. Each of the other 2 divide at the clefts distally to supply the opposing halves of the index and the middle, and the middle and the ring fingers. Motor branches to the lumbricals of the index and the middle fingers are given off from these digital nerves. (The ulnar nerve supplies the other 2 lumbricals, all the interossei, the adductor pollicis and the deep head of the flexor pollicis brevis.) The motor and the sensory distributions of the medial and the ulnar nerves frequently overlap. Occasionally, even the opponens may be supplied by the ulnar. Normally, the median nerve supplies the palmar surface of the thumb, the index, the middle and the radial half of the ring fingers and the dorsal surface of the distal thirds of these fingers. The distal ends of the index and the middle fingers volar and dorsal invariably have no overlap of sensory supply from radial or ulnar nerves, and absence of sensation in this area in median nerve severance is usually complete.

Findings After Median Nerve Injury.

Severance above the elbow results in loss of flexion of the thumb, the index and the middle fingers; wrist flexion is weak and deviates ulnarward from unopposed action of the flexor carpi ulnaris; pronation is weak or absent; the thumb is in a position at the side of the hand and cannot be brought forward into a position of opposition; the upper forearm and the thenar area lose their normal convexity because of atrophy; loss of sensation is experienced in the volar aspect of thumb, index, middle and radial half of ring fingers. Occasionally, the opponens function may be intact because of anomalous ulnar nerve supply, but this function is generally inadequate in that the thumb cannot be rotated so that its nail is parallel with the palm. The appearance of the hand is similar to the flat hand of the monkey and therefore is called "simian hand." Trophic disturbances occur chiefly at the distal end of the index finger, which becomes thin and conical. Injuries of the median nerve above the elbow, in addition to severance by missiles or sharp instruments, may result from the jagged edges of a supracondylar fracture.

Lesions at the wrist occur frequently from accidental cuts by knives or broken dishes or suicide attempts. The nerve may be compressed against the nonyielding transverse carpal ligament by a dislocated semilunar bone or by strongly grasping an object, particularly with the wrist in flexion whereby the flexor tendons are strongly displaced volarward. Rarely, a condition of abnormal thickening of the transverse carpal ligament can reduce the caliber of its subjacent canal, thereby constricting the nerve. A lesion at the wrist level is beyond the level of supply to the long

flexors of the thumb, the index and the middle fingers, the radial carpal flexor and the pronators. The paralysis affects the short abductor (inability to bring the thumb far forward opposite the index finger), the opponens, the superficial part of the short flexor, and the lumbricals to index and middle fingers. The loss of sensation is the same as in higher lesions.

Partial nerve injury or irritation of the median nerve is the most common cause of *causalgia*. This is characterized by severe burning pain in the extremity, especially the hand, aggravated by physical or emotional stimuli. The hand initially may be swollen, red, warm, perspiring, hyperesthetic. Gradually, the skin becomes thinned, glossy, cold, cyanotic and dry. The hand is held fixed with the fingers extended and the thumb adducted, and the joints may ankylose in this position. Pain becomes extremely distressing. Keeping the part moist seems to reduce the symptoms temporarily.

Surgical Treatment. Decision should be made as to whether conservative or surgical treatment is to be done. In nerve suture, better results obtain from early intervention. Late suture generally leads to partial restoration, particularly of sensation, and paresthesias. The nerve should be explored, and the ends obtained and sutured in exact rotary apposition. Gaps between nerve ends can be overcome in the palm by flexing the metacarpophalangeal joint. Above the wrist the nerve is freed, and the wrist is flexed. If the elbow is also flexed and the nerve is gently pulled distally, a 3½-inch gap can be overcome. For larger gaps, it is necessary to dissect the nerve in the upper arm and reroute it superficial to the elbow structures by detaching the humeral head of the pronator teres. A plaster cast maintains flexion of the joints, and very gradual extension is obtained over a period of 1 month. If a graft is needed, the sural nerve may be used. This extensive surgery is justified by the serious disability caused in workers by loss of the important tactile sense in the median nerve area. If opposition is inadequate after nerve suture, the movement may be restored by a pulley operation around the insertion of the flexor carpi ulnaris.

At exploratory operation in causalgic states,[17] the nerve displays a lesion in continuity, i.e., intraneural scarring. Complete division of a nerve rarely causes causalgia. In wartime series, high velocity missiles or bomb splinters were the main causes, the injury practically always being above the elbow in case of the median nerve (above the knee in the lower extremities). Treatment consists of sympathectomy. This is preceded by procaine block of the second thoracic ganglion. Complete anhidrosis and increase in warmth of the hand appearing 10 minutes after the block demonstrates effectiveness of the block. Pain is relieved for 1 to 3 hours. A preganglionic sympathectomy is most effective. The white rami communicantes to the 2nd and the 3rd thoracic ganglia and the sympathetic trunk below the third are divided.

Radial Nerve

Anatomy. The radial nerve is the continuation of the posterior cord which is formed by the posterior divisions of the brachial plexus In the axilla it lies directly behind the axillary artery and with the other neurovascular structures runs on a floor formed by the subscapularis muscle proximally and the latissimus dorsi and the teres major distally. The axillary (circumflex) nerve, which originates from the posterior cord, descends alongside the radial nerve, then leaves it at the lower border of the subscapularis, where it passes backward through the quadrangular space.

Beyond the teres major, the radial nerve proceeds posterior to the humerus by entering an interval between the long and the medial heads of the triceps and reaching the spiral groove between the medial and the lateral heads of the muscle. It passes around the back of the humerus to the lateral side, where it pierces the lateral intermuscular septum to reach the anterior aspect of the arm. Here it lies in an interval between the brachialis medially and the brachioradialis and the extensor carpi radialis longus laterally. At this level it gives off branches of supply to the lateral half of the brachialis, to all of the brachioradialis and the extensor carpi radialis longus and to the posterior interosseous nerve.

[17] Barnes, R: The role of sympathectomy in the treatment of causalgia, J Bone & Joint Surg. 35B: 172, 1953.

It then continues distally in the forearm under cover of the brachioradialis until a level about 2 inches above the wrist is reached. Here it pierces the deep fascia and turns laterally and dorsally, crossing superficial to the tendons of the long abductor and the short extensor of the thumb, gaining the dorsum of the hand where it supplies digital branches of sensation to the dorsum of the thumb, the index, the middle and the radial half of the ring fingers as far as the middle phalanges. In the spiral groove the radial nerve gives off the posterior cutaneous and the lower lateral cutaneous nerves of the arm, the posterior cutaneous nerve of the forearm, and muscular branches to the triceps and the anconeus.

The *posterior interosseous nerve* springs from the radial nerve at the level of the lateral epicondyle. It descends under cover of the brachioradialis, gives branches to the extensor carpi radialis brevis and the supinator. Then it penetrates the supinator and passes obliquely around the lateral aspect of the shaft of the radius to reach the back of the forearm and travels distally on the surface of the abductor pollicis longus and under cover of the extensor digitorum. Then it lies on the interosseous membrane under cover of the extensor pollicis longus and proceeds distally to supply the wrist joint. In the back of the forearm it supplies the remainder of the extensor muscles and the abductor pollicis longus. Therefore, it supplies all muscles on the lateral and the dorsal aspects of the forearm except the brachioradialis and the extensor carpi radialis longus, which are supplied directly by the radial nerve.

Clinical Picture. The extent of motor and sensory findings depends on the level of the injury and the degree of trauma. When the radial nerve is interrupted at the axilla where it is usually involved by direct compression, such as by the arm resting over the back of a chair (Saturday night palsy) or by pressure of a crutch (crutch palsy), the extensors of the elbow, the extensors and the supinators of the forearm, the extensors of the wrist, the extensors of the MP joints of the fingers, and the extensors and the long abductor of the thumb are paralyzed. A strip of the posterior and the posterolateral surface of the arm, the posterior third of the forearm and an autonomous zone on the dorsum of the hand over the first interosseous space are anesthetic.

The typical picture is one where the patient holds the extremity at the side, the elbow is slightly flexed, the forearm is pronated, the hand is dropped at the wrist, and the fingers are dropped at the MP joints. The thumb is turned forward into the palm and interferes with flexion of the fingers. The patient cannot make a fist because the drop wrist tenses the extensors of the fingers and thereby opposes their flexion.

Involvement of the nerve in the spiral groove may be immediate and caused by the sharp jagged edge of a fracture fragment; or it may be delayed by formation of callus about and incarceration of the nerve. An injury to the nerve beyond the spiral groove permits the function of the triceps and the anconeus and preserves sensation at the back of the arm and the forearm. The autonomous area at the back of the hand supplied by the superficial radial nerve is anesthetic.

Injury to the radial nerve in the interval between the brachioradialis and the brachialis involves the brachioradialis and the extensor carpi radialis longus. The brachialis, which has a dual nerve supply, continues to function. The autonomous sensory area of the hand is affected. Gunshot and stab wounds are main offenders at this level. These muscles escape, and hand sensation is preserved when the injury is at the level of the radius, where the posterior interosseous nerve encircles the bone one fingerbreadth below the head of the radius. Surgical trauma is the frequent etiologic agent. Beyond this level, the supinator brevis is permitted to function, whereas a wrist-drop, a finger-drop at the MP joints, and the thumb rolled forward into the palm are the deformities. The thumb extensors and the long abductor gain their branches of supply a little more distally than the extensor digitorum communis and the wrist extensors, so it is possible to have the thumb alone involved by a properly placed point of trauma.

Missiles and stab wounds produce most nerve injuries in the forearm. When the superficial radial nerve alone is severed, the loss is restricted to sensation in the autonomous zone. This area is the main site of pain when a causalgic state results from incomplete

lesions of the superficial radial. Partial paralysis of one or several muscles and hypesthesia or hyperesthesia rather than anesthesia indicate that the nerve lesion is incomplete and continuity of the nerve is preserved.

Automatic movements at the wrist should not be interpreted as preservation of the wrist extensors. When the fingers are flexed, the extensor tendons are tightened, and the hand is drawn backward at the wrist.

Examination of extension of the fingers should be directed to the MP joints. The lumbrical muscles supplied by the median and the ulnar nerves extend the distal two phalanges at the interphalangeal joints.

By force of gravity, the elbow is extended in spite of triceps paralysis. Examination is performed by keeping the arm horizontal and the forearm dependent and attempting to extend the elbow actively, or the entire extremity may be supported on a flat surface with the elbow flexed, and active extension is attempted.

Treatment. Regardless of the level or the cause of nerve injury, the affected muscles should be kept in a state of relaxation by supportive splints and their tone maintained by galvanic stimulation and light massage until the nerve regenerates An anterior molded splint counteracts the drop wrist and should extend beyond the MP joints to support the proximal phalanges. An additional extension from the splint holds the thumb in complete extension and dorsal abduction.

If paralysis is immediate and complete, the nerve should be explored and sutured promptly. Good results are proportionate to early repair. Nothing is lost by venturing an early exploratory operation and finding the nerve intact.

Gaps between nerve ends may be overcome by flexing the elbow, by externally rotating and adducting the arm and freeing various branches. If the distance is extensive, occasionally the nerve may be transposed anteriorly. Shortening of the humerus is sometimes justified to aid approximation.

Compression injuries, no matter how extensively the muscle is involved, are generally temporary, and almost complete restoration of function is the rule. Various texts also include in this category radial nerve injuries due to fractures at the middle third of the humerus. However, the possibility of complete nerve tears and their serious implications certainly warrants operative exposure of the fracture site, whereupon both nerve and bone injuries can be dealt with at the same time.

In situations of indecision as to whether to operate, the muscles should be studied daily by electric reaction tests. If the galvanic response seems to be disappearing, loss of nerve continuity is highly probable. Certainly, failure of muscles to recover spontaneously during the initial period of 8 to 10 weeks is justification for exploring the nerve.

When a causalgic state arises in the distribution of the radial nerve, an incomplete nerve lesion should be suspected, the nerve explored, and the pathology dealt with. A neuroma should be resected if necessary by removal of a portion of the nerve, followed by reapproximation; or, adhesions may be freed and the nerve surrounded by fatty tissue or paratenon or imbedded in adjacent muscle. The Tinel sign may reveal the exact site of initiation of pain impulses.

Prognosis. The prognosis in radial nerve repair is usually very good. However, failure of some portion to regenerate necessitates tendon transplantation. Triceps paralysis needs no compensation other than that provided by gravity. Extension at the elbow is rarely ever required. Such an instance is the need for use of crutches, whereupon the brachioradialis origin displaced to a more posterior site on the humerus will provide satisfactory elbow extensor power.

Supination of the forearm may be restored by osteotomy of the radius and rotating the distal fragment. The Tubby operation transplants the insertion of the pronator teres from the volar to the dorsal aspect of the radius. Extension of the thumb and particularly abduction which stabilizes the digit at the carpometacarpal joint is necessary for proper apposition of the thumb to the fingers in the functions of pinch and grasp. The flexor carpi radialis may be transplanted to the long abductor and both extensors. The flexor carpi ulnaris is transferred to the finger extensors.

If no tendons are available for transference, dorsiflexion of the wrist is provided either by arthrodesis of the wrist or by severing the tendons of the extensor communis and tenodesing the proximal ends of the distal seg-

ments to the dorsum of the radius. Active flexion at the MP joints thereby tightens these tenodesed tendons and automatically dorsiflexes the wrist. The carpometacarpal joint of the thumb may also be stabilized by arthrodesis.

Ulnar Nerve

Anatomy. The ulnar nerve is the largest branch of the medial cord of the brachial plexus, arising under cover of the pectoralis minor and descending along the medial side of the axillary artery and the proximal half of the brachial artery. At the level of insertion of the coracobrachialis at the middle of the humerus, it leaves the brachial artery and, in company with the ulnar collateral artery, it passes backward through the medial intermuscular septum to the posterior aspect of the arm. Then it descends along the medial head of the triceps to the back of the medial epicondyle and passes between the heads of the flexor carpi ulnaris to enter the forearm. There, under cover of the flexor carpi ulnaris (which it supplies), it lies upon the flexor digitorum profundus (supplies its medial half) and is immediately lateral to the ulnar artery.

Near the pisiform bone it emerges through the deep fascia lateral to the flexor carpi ulnaris and descends anterior to the flexor retinaculam, where it divides into superficial and deep branches. The deep branch passes medial to the hook of the hamate and, with the deep branch of the ulnar artery, enters the interval between the abductor and the flexor of the little finger to gain the deep area of the palm. It gives off branches of supply to the hypothenar muscles, then turns laterally across the palm deep to the flexor tendons, giving off 3 branches, each of which runs distally in front of the interosseous space, supplying the interosseous muscles. The medial 2 branches also supply the medial 2 lumbrical muscles. At the lateral side of the palm, the main deep branch of the ulnar nerve ends by breaking up into nerves of supply to the adductor pollicis and the first dorsal interosseous muscle. The superficial branch of the ulnar nerve runs under the palmaris brevis, which it supplies, then divides into 2 digital branches which provide sensation to the palmar aspect of the little finger and the ulnar half of the ring finger.

The dorsal branch of the ulnar nerve arises from the latter at the middle of the forearm and descends with the parent nerve to the carpus, where it becomes superficial and inclines backward to gain the dorsum of the hand. Here it divides into 2 dorsal digital nerves which supply the skin of the medial third of the back of the hand and the little finger and the ulnar half of the ring finger as far as the second phalanx.

Clinical Picture. When the flexor carpi ulnaris is paralyzed (on attempting flexion at the wrist, the hand deviates radialward), interruption of the ulnar nerve has occurred above the elbow. Preservation of this muscle's function places the lesion distal to the elbow. Otherwise, ulnar nerve paralysis is typified by the following: The ring and little fingers are extended at the metacarpophalangeal joints and flexed at the proximal interphalangeal joints, because of loss of lumbrical action. When the lesion is sufficiently low in the forearm, the flexor profundus is spared and, unopposed by the intrinsics, exerts strong flexion on the distal phalanges, and clawing of the ring and the little fingers is pronounced.

When the flexor carpi ulnaris and the ulnar portion of the flexor profundus are paralyzed by a high lesion, the ensuing atrophy over the ulnar aspect of the forearm is very apparent. In this instance, flexor power to the distal phalanges of the ring and the little fingers is lost. This is demonstrated best by placing the hand palm down when the inability of the little finger to scratch the surface of the table is evident. The hypothenar eminence is thinned, and the hollowing of the interosseous spaces attests to atrophy of the paralyzed interossei, the thumb adductors, the inner head of the flexor pollicis brevis, the hypothenars and the two lumbricals on the ulnar side. Abduction and adducion of the fingers is lost to a great extent. The index and the middle fingers may still abduct because their lumbrical innervation through the median is still intact.

Pinch between the thumb and the index finger normally is dependent upon the ability to stabilize the metacarpophalangeal joint in flexion (adductors and flexor brevis) so that such action is strong and the apposed fingers form the letter O. In ulnar paralysis, the proximal phalanx becomes hyperextended, the in-

terphalangeal joint of the thumb hyperflexes, and the pinch is weak. Failure of stabilization at the carpometacarpal joint by the abductor pollicis longus will likewise interfere with pinch. Loss of thumb adductors is demonstrated by inability of the thumb to scrape across the distal palm. Instead, it comes forward into the opposed position. Another test is failure to resist attempts to extract a sheet of paper held between the apposed sides of the thumb and index finger. On the volar aspect, sensation is lost over the ulnar portion of the hand and all of the little finger and the ulnar half of the ring finger. On the dorsum, the entire little finger, the ring finger and the ulnar half of the long finger and the ulnar third of the hand are involved. However, because of overlap from adjacent nerves, loss of sensation is variable, but the distal two thirds of the little finger are independent and invariably involved.

Trophic changes in the ring and the little fingers reflect the loss of sensory innervation. Very frequently innervation from the median nerve preserves function of the intrinsics and the thumb adductors. Fibers innervating these muscles may proceed distally in the median nerve to the distal third of the forearm and by a connecting branch enter the ulnar nerve before the latter reaches the hand. In such an instance, a high ulnar lesion fails to eliminate intrinsic action.

Treatment. Repair of the ulnar nerve should be done with care. The nerve contains both motor and sensory fibers, and accurate coaptation will prevent sensory fibers from growing down motor pathways, and vice versa. Gaps are overcome by flexing the wrist and the elbow. The nerve at the elbow may be transposed anteriorly, and branches are freed, thereby permitting mobilization distally. Recovery of function requires more than a year and occurs in the following order: forearm muscles, sensation, hypothenar muscles, interossei and thumb adductors. During this long recovery period, the hand is splinted with the metacarpophalangeal joints in flexion and the interphalangeal joints in extension to keep the paralyzed muscles in a relaxed state and prevent joint contractures. A stretched muscle will become fibrotic and void the result of nerve suture.

When ulnar nerve paralysis is permanent, tendon transplantation is the treatment of choice. Necessary prerequisites are mobile joints and good muscles for transfer.

Surgical Repair of Clawed Fingers.[15] A clawhand deformity is due to intrinsic muscle paralysis while the long extensors and the long flexors are still functioning. The loss of flexor power on the proximal phalanges allows the extensors to pull the proximal phalanges into hyperextension; tension on the long flexors pulls the distal phalanges into flexion, unopposed by the lost extension of the intrinsics.

Extension of the distal 2 phalanges takes place synergistically by the long extensors and the intrinsics (Fowler). The action of the long extensors is lost when the proximal phalanx is hyperextended. Any procedure which prevents hyperextension of the proximal phalanx preserves extension of the distal 2 phalanges and eliminates the claw deformity. The following are the most commonly used procedures:

Bunnell Technic. This operation transplants multiple slips of sublimis tendons through the lumbrical canals into the aponeurotic expansion. This procedure is not effective in clawhand of long standing where the patient has developed the habit of flexing the wrist to extend the distal phalanges automatically, thereby rendering the sublimis impotent.

Fowler Technic. This procedure splits the extensor indicis proprius and the extensor digiti quinti into 2 strands each; next, each individual slip is passed through the interosseous space anterior to the transverse metacarpal ligament and inserted into the aponeurosis.

Riordan Technic. This is a tenodesis procedure. Half of the extensor carpi radialis longus and the extensor carpi ulnaris is separated from the parent tendon and left attached to the insertion into the base of the 2nd and the 5th metacarpals respectively. Each half is split longitudinally into 2 slips, and then each slip is passed and attached as in the Fowler operation. The tendon should be under tension at the conclusion of the operation so as to obtain restriction of extension.

Postoperatively, the hand is immobilized in a pressure dressing, maintaining the wrist in

[15] Riordan, D. C.: Tendon transplantation in median and ulnar nerve paralysis, J Bone & Joint Surg. 35A:312, 1953.

dorsiflexion, the metacarpophalangeal joints in flexion, and the distal 2 joints in extension. Tendon transplantation is unsuitable in deformity with skin and joint contractures. These require joint arthrodeses.

Restoration of Thumb Adduction and Carpal and Metacarpal Arches. When grasping small round objects, the hand cups into an arch and enables the fingers to converge during flexion, and strength of grasp is obtained. These arches are produced mainly by the thenar and the hypothenar muscles and are reduced considerably in ulnar paralysis and are completely flattened in combined median and ulnar paralysis. It is essential to restore the functions of pinch and grasp. The following procedures are used:

1. *Tendon Loop Operation.* The tendon of the extensor communis to the index finger is removed just before its insertion and is prolonged by a tendon graft around the ulnar border of the hand, placed volar to the hypothenars and beneath the finger flexors and inserted into the ulnar side of the base of the proximal phalanx of the thumb. The distal remaining stump of the tendon is attached to the extensor indicis to avoid adduction and rotation deformity of the index finger. This procedure restores only adduction and is suitable only in pure ulnar paralysis.

2. *Tendon T Operation* This provides strong adduction to both thumb and index fingers and reforms the carpal and the metacarpal arches. A tendon graft is placed transversely across the palm beneath the flexor tendons and is attached to the neck of the 5th metacarpal and the ulnar side of the base of the proximal phalanx of the thumb. To its center is attached a motor tendon, usually one of the sublimis tendons. Contraction of the motor tendon pulls on the cross member and apposes the thumb and the index finger. This procedure is done when median nerve paralysis is associated with ulnar involvement and it is necessary to correct thumb opposition also These problems are discussed more fully in the section on "The Hand"



Fig. 140. Cubitus valgus, the result of malunion of fracture at the lower end of the humerus. Degenerative arthritis has supervened. Stretching of the ulnar nerve over the medial prominence is a common sequel.

Traumatic Ulnar Neuritis.[19] The ulnar nerve is most commonly subjected to chronic repeated traumata where it is most superficial at the groove between the olecranon and the medial epicondyle of the humerus. Here it is subjected to external pressure, as in occupational situations. The nerve is held firmly against the subjacent bone by a strong dense fascia so that roughening of the bone produced by osteoarthritis or previous fracture imposes constant friction on the gliding nerve. When a cubitus valgus deformity exists, the nerve is supposedly stretched over the medial prominence of the elbow and results in ulnar nerve symptoms. The valgus may be congenital and associated with anterior dislocation of the radial head, or it may be acquired following fracture through the outer aspect of the epiphyseal plate and reduced growth rate. A

[19] McGowan, A J. Results of transposition of the ulnar nerve in traumatic ulnar neuritis, J. Bone & Joint Surg. 32B 293, 1950.

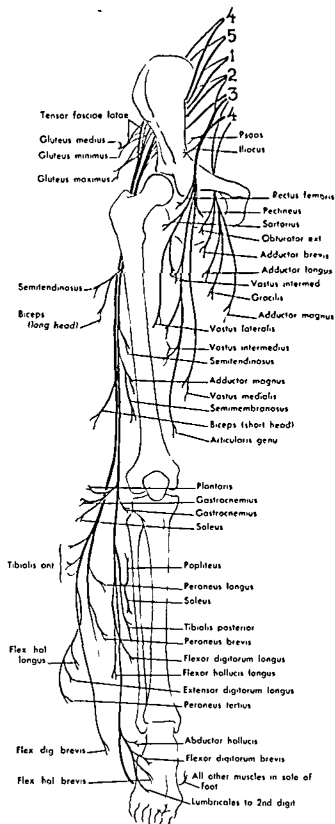


FIG. 141. Nerve supply to muscles of the lower extremity.

shallow groove or inability of the overlying fascia to retain the nerve in place allows recurrent dislocation of the nerve. Scarring as a result of direct trauma or infection can constrict the nerve.

Minimal irritation of the nerve causes paresthesias and slight blunting of sensation in the ulnar distribution, feeling of clumsiness of the hand, and occasional hyperhydrosis. Grossly, the nerve appears to be normal or is surrounded by a few filamentous adhesions. Moderate involvement of the nerve causes more intensive symptoms and findings such as interosseous weakness and wasting, hypothenar and forearm muscles weakened but to a lesser degree than the intrinsics, hypesthesia to anesthesia, and preservation of sweating, occasionally hyperhydrosis. The nerve displays moderate swelling or a fusiform neuroma with enveloping adhesions or compression by scar tissue. Infrequently, the nerve has a normal appearance. Severe lesions are characterized by more marked involvement. The interossei are very weak or paralyzed and atrophied. The hypothenars, the flexor carpi ulnaris and the ulnar half of the flexor digitorum profundus are partially weakened and atrophied. Sensation varies from marked hypesthesia to analgesia and anesthesia in the ulnar area. Sweating is generally reduced, although hyperhydrosis is not infrequent. A large fusiform neuroma or swollen nerve trunk is found.

Treatment. This condition is treated by removing the nerve from its groove and transposing it to a position anteriorly. The fascia over the nerve is incised, the medial intermuscular septum is resected, the nerve is placed superficial to the flexor muscles, and the superficial fascia is sewed to the deep fascia by one stitch to prevent the nerve from redisplacing backward. Occasionally, when the neuroma is very large and tender, or scarring is excessive over the medial elbow, or too little fatty covering is available, the nerve is transposed deep to the flexors. This is done by removing the origin of the common flexor tendons at the medial epicondyle, displacing the nerve beneath the flexors anteriorly, and replacing the tendon origin. The earliest and most constant result is relief of discomfort and ulnar paresthesia. The degree of motor recovery varies with the severity of the lesion. Recovery of sensation is uniformly good.

SCIATIC NERVE AND SCIATICA

Anatomy. Sciatica is the term applied to the condition of pain in the area of distribution of the sciatic nerve. It is due to a large

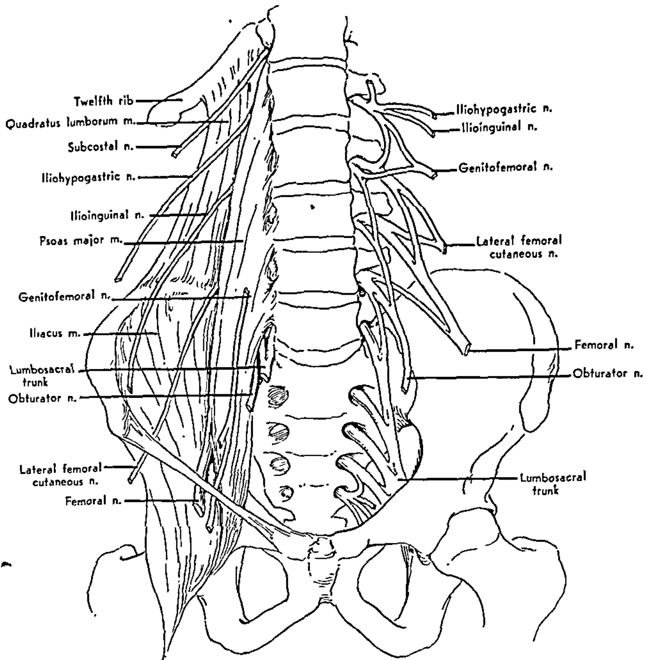


FIG. 142. Lumbosacral plexus. Note the position of individual nerves as they emerge from under cover of the psoas major muscle. Involvement of these nerves by visceral disease in the lumbar region is reflected in sensory disturbances in the lower abdominal, the inguinal and the lateral femoral areas. Involvement by pelvic disease refers dysesthesias to the anterior femoral area (femoral nerve) and the posterior aspect of the entire lower extremity (lumbosacral trunk).

variety of causes which are intraspinal, intrapelvic and extrapelvic. Therefore, knowledge of the anatomy is a necessary prerequisite to making a differential diagnosis.

The sciatic nerve arises from the 4th and the 5th lumbar and the 1st, the 2nd and the 3rd sacral nerves. These nerve roots, after emergence from the spine, form the sacral plexus within the pelvis. The 4th and the 5th lumbar nerves form the large lumbosacral trunk which appears at the medial margin of the psoas major and descends downward over the pelvis brim to join the 1st sacral nerve.

The plexus lies on the posterior wall of the pelvis between the piriformis behind and the hypogastric vessels, the ureter and the sigmoid colon in front. The plexus gives rise to nerves to the gluteal muscles and the external rotator

muscles of the hip, then converges distally toward the greater sciatic foramen as a large flattened band, the sciatic nerve.

The nerve emerges from the foramen at the lower border of the piriformis and comes to lie between the other external rotators anteriorly and the gluteus maximus posteriorly. The rotators and the nerve are situated in a groove between the ischial tuberosity medially and the greater trochanter laterally. At this point it is exposed to pressure from the posteriorly displacing femoral head in dislocations.

The sciatic nerve descends beneath the obliquely placed biceps femoris long head and lies on the adductor magnus, very close to the back of the femur. In this position it is easily compressed between the bone and a hard chair seat and gives rise to the sensation of the foot falling asleep; or it may be involved in tumor or callus formation. Sharp jagged edges of fragments may sever the nerve. Branches springing from the sciatic supply the hamstrings and the posterior part of the adductor magnus. Nerves of supply to the biceps and the semitendinosus arise quite high so that these muscles are generally spared in lesions of the sciatic, and knee flexion is preserved. A lower branch to the semitendinosus and a common branch for the adductor and the semimembranosus complete the distribution in the thigh. In the lower third of the thigh, the sciatic divides into the tibial (medial popliteal) and the common peroneal (lateral popliteal) nerves.

The tibial nerve in the popliteal space lies to the lateral side of the large vessels, then crosses over them and comes to lie medially In the lower fossa, branches are given off to both heads of the gastrocnemius and to the soleus, the popliteus and the plantaris. A cutaneous nerve, the sural, is given off here and descends superficially in the calf to the lateral side of the foot and the little toe. At the lower end of the popliteal fossa, it exits through the tendinous arch in the soleus and descends in the leg under cover of that muscle, lying on a floor formed by the posterior tibial and the flexor digitorum longus muscles.

At the lower third of the leg it lies in intimate contact with the posteromedial surface of the tibia, where it may be easily affected by bony lesions. Then it passes under the flexor retinaculum behind the medial malleolus and enters the sole of the foot, where it divides into mediál and lateral plantar nerves. These supply the small muscles to the toes and the skin of the sole except the extreme medial and lateral borders.

The tibial nerve, throughout its course in the leg, lies just behind and lateral to the tibial vessels and together with the latter are held in intimate contact with the deep calf muscles by an enveloping layer of fascia. At the level of the medial malleolus, the neurovascular bundle lies one fingerbreadth posterior to the malleolus.

The common peroneal nerve originates at the upper end of the popliteal fossa and passes distally and laterally toward the head of the fibula in close relationship to the posterior border of the biceps tendon. At the level of the fibular head it lies in a groove between the head, to which the biceps inserts, and the gastroenemius. Here it gives off the lateral sural cutaneous nerve, which supplies the skin on the posterior and the lateral surfaces of the leg. Then it passes around the neck of the fibula where, under cover of the origin of the peroneus longus, it divides into the superficial and the deep peroneal nerves. These branches are in close contact with the bone and are easily damaged. Surgery in this area demands mobilization of the nerves by first securing the common peroneal above and behind the biceps, then loosening it distalward. Finally, by severing the posterior attachment of the peroneus longus, the main nerve and the branches may be displaced forward with the muscle out of harm's way.

The deep peroneal nerve passes obliquely forward beneath the extensor digitorum longus to the front of the interosseous membrane and comes into intimate relationship with the anterior tibial artery with which it descends distally to the front of the ankle joint. Here it divides into medial and lateral terminal branches; the former supplies the dorsal surfaces of the apposing aspects of the large and the 2nd toes; the latter innervates the extensor digitorum brevis. In the leg, the deep peroneal supplies the anterior tibial, the extensor hallucis and the extensor digitorum longus muscles at a high level, so that a lesion in the distal portion of the leg affects the foot extensor power very little. Above the ankle, the neurovascular bundle is found in the in-

terval between the tibialis anticus and the extensor digitorum longus.

The superficial peroneal nerve originates about the neck of the fibula and innervates the long and the short peroneal muscles as it descends distally in close contact with the bone. Then it runs superficial to the brevis and passes obliquely forward between the peronei and the extensor digitorum longus and divides into a medial dorsal cutaneous nerve and an intermediate dorsal cutaneous nerve. The former supplies the skin on the medial side of the great toe and the adjacent sides of the 2nd and the 3rd toes; the latter supplies the adjacent sides of the 3rd and the 4th toes and the 4th and 5th toes. The lateral side of the 5th toe is supplied by the sural.

Clinical Picture of Sciatic Nerve Involvement. Sciatica, defined, is pain in the area of distribution of the sciatic nerve, i.e., over the buttock, the posterior thigh, the posterior or postero-lateral leg, and the foot, dorsal or plantar. The following are the most important causes of sciatic nerve pain:

1. Primary Nerve Disease
   - A. *Inflammatory*—alcoholic, avitaminosis, infection (spread from pelvis, hip, etc.)
   - B. *Degenerative*—heavy metals
   - C. *Metabolic*—diabetes
2. Secondary Nerve Involvement
   - A. *Intraspinal*
     - a. Intervertebral disk compression of nerve root
     - b. Hypertrophic arthritis
       - (1) Osteophyte compression
       - (2) Edema at foramina
     - c. Post-traumatic arachnoiditis, fracture
     - d. Vertebral disease
       - (1) Tumor
       - (2) Infection
     - e. Intraspinal tumor
     - f. Congenital deformities—spondylolisthesis
   - B. *Extraspinal*
     - a. Intrapelvic
       - (1) Tumors, e.g., prostate
       - (2) Infection, e.g., postabortive
     - b. Extrapelvic
       - (1) Trauma, e.g., gunshot, dislocated hip
       - (2) Tumors, e.g., pressure of osteochondroma
       - (3) Infection
3. Reflex Nerve Pain
   - A. Tumors, e.g., episacral lipoma, osteoid osteoma
   - B. Trauma, particularly low back strains.

These by no means are all the causes of sciatica, but the outline is designed to clarify for differential diagnosis.

The sciatic nerve carries not only the described motor and sensory fibers but also sympathetic fibers. Each motor supply is to a definite muscle or group of muscles. The sensory fibers supply a definite area also, and in spite of overlap from adjacent peripheral nerves there is an autonomous zone of sensory loss that defines the particular nerve involvement. When the peripheral nerve is irritated, pain of a constant burning type in the area of distribution results. Also, the stimulation of sympathetics causes vasoconstriction (pallor) and increased sweating in the autonomous area. When the nerve is injured further so that function is lost, in addition to paralysis there results anesthesia in the autonomous zone, increased warmth (vasodilatation) and dryness. Trophic lesions are common. Next to the median nerve, the sciatic is a frequent site for causalgia.

Lesions proximal to the lumbar plexus, i.e., at the level of the nerve roots, causes an incomplete sensory loss corresponding to a dermatome or segment. Muscle weakness or paralysis involves a group of muscle fibers, not necessarily anatomically distinct muscles, corresponding to a myotome. No vasomotor or sudomotor functions are lost. This picture contrasts sharply with peripheral nerve lesions. Thus, the clinical picture of a disk compressing a nerve root at the lumbosacral interval may display hypesthesia over the lateral lower leg, slight weakness of dorsiflexion at the ankle when attempted against resistance, and no change in color, warmth, or moisture in the skin.

When the sciatic nerve is inflamed, pressure over the nerve by the palpating finger will accentuate the pain. The nerve is most accessible to examination at (1) the groove between the greater trochanter and the ischial tuberosity;

(2) the popliteal space; (3) at the posterior edge of the biceps tendon; (4) and at the neck of the fibula. Any stretching of the nerve, as when performing the Lasegue maneuver, will increase the pain when the nerve is inflamed, so that the test does not necessarily point to a low back lesion. When the test is positive in the absence of nerve tenderness, it suggests that the nerve is displaced over a prominence, for example, a disk or a bone tumor.

Differential Diagnosis. The differential diagnosis of sciatica should begin at the proximal areas and proceed distally. The back is inspected for deformities and abnormal posture. Inequalities in length of the lower extremities are determined. Evidence of hip dislocation and instability should be sought. The soft tissues about the lower back, particularly at the posterior superior iliac spine, are examined for tender lipomatous masses. Local anesthesia of a suspected mass will temporarily relieve reflex sciatic pain if this be the cause. A roentgenogram of the spine demonstrates bony lesions and narrowing of disk spaces. One should remember that narrowing of the disk space and concomitant degenerative arthritic changes about the facets at the same level are late results of a disk long since gone by rupture or degeneration Given such a situation, compression of a nerve root by disk protrusion is unlikely. Oblique views of the lumbar spine will show interruption in continuity of the pars interarticularis characteristic of spondylolysis and spondylolisthesis; encroachment on the foramina by osteophytes can be seen Pantopaque myelography may display defects characteristic of various space-occupying lesions. Spinal fluid studies give further information such as the increased protein content of tumors and disks, manometric evidence of a spinal block, and tests for syphilis.

The abdomen is examined for evidence of lesions which by their involvement of the lumbosacral plexus could produce sciatic pain The male genito-urinary system may reveal a prostatic tumor which by spread backward could encompass the nerve trunks Similarly, the tumors peculiar to the female reproductive system, such as endometriosis, are causative. Bony lesions in the pelvis may compromise the nerve before or after it emerges from the greater sciatic foramen. At the level of the hip, the most common causes are traumatic posterior dislocation and the large osteochondral growths peculiar to this region. Beyond this site, fractures of the femoral shaft may injure the nerve. Direct compression of the nerve against the unyielding underlying bone may entirely destroy nerve function, or irritative lesions will result in the causalgic state. At the neck of the fibula, the peroneal nerve is affected by compression, as by a cast, or improper surgical approaches. The investigation always should include thorough roentgenography of the entire extremity. An unsuspected bony tumor, such as an osteoid osteoma, which exerts its influence reflexly, may thereby be revealed.

/ When the sciatic nerve is interrupted in the thigh, usually the ability to flex the knee is not lost, because the biceps femoris and the semitendinosis are innervated very high. A flaccid dangling foot which is anesthetic results. Anesthesia extends to the posterior and the lateral aspects of the leg. When the common peroneal nerve is involved, the ability to dorsiflex the foot and the toes and to evert the foot is lost. Sensory loss is found over the dorsum of the foot and the toes except the lateral side of the little toe. The superficial peroneal nerve injury results in loss of eversion of the foot and reduced sensation over the medial aspect of the big toe and the apposing surfaces of the 2nd and the 3rd, the 3rd and the 4th toes, and the 4th and the 5th toes. The deep peroneal (anterior tibial) nerve must be injured high in the leg in order that innervation to the foot and toe extensors be interrupted. Otherwise, the only effect is loss of sensation between the large and the 2nd toes.

In contrast with the peroneal nerve, which is the most exposed of the sciatic components, the tibial nerve is rarely injured. Its loss results in paralysis of the calf and the plantar muscles and their obvious atrophy. Peculiarly enough, the gait is little disturbed, and the functional defect becomes apparent only when fast walking or running is attempted; then loss of take-off is demonstrable. Clawfoot deformity results from the unopposed pull of the extensors at the metacarpophalangeal joints, because of paralysis of the interossei and the lumbricals. The loss of extension at the interphalangeal joints by these

muscles causes flexion deformity of the toes. The deformity is comparable with that seen in the hand. The sensory loss includes the sole of the foot and the plantar aspect of the toes. If the sural nerve is also involved, the posterolateral leg and the lateral border of the foot and little toe are also anesthetic.

The autonomous zones should be sought and outlined by the starch-iodine test or by determining the skin resistance by an instrument such as the Richter dermometer. In the case of the sciatic nerve, the autonomous zone includes the entire dorsum and plantar surfaces of the foot, except a small medial area, and the lower lateral aspect of the leg. The autonomous zone of the peroneal is extremely variable over the dorsum of the ankle and the foot. The zone for the tibial nerve exists over the sole of the foot and the toes and the lateral surface of the heel.

Surgical Repair of Sciatic Nerve Lesions. Because of the great length of the sciatic nerve, regeneration is prolonged, and repair should be undertaken at the earliest possible date. By percussion along the nerve, the site of injury is determined by the painful tingling sensation produced.

APPROACH TO THE SCIATIC NERVE. The incision begins at the posterior superior iliac spine and is carried obliquely downward and outward to a point medial to the great trochanter. Then it is curved medially at the gluteal fold to the mid-line of the thigh and then continued distally to a point just above the popliteal fossa. At the upper end of the incision, the gluteus maximus is split in line with its fibers, and laterally it is severed from its insertion into the iliotibial band. The muscle is reflected medially, exposing the nerve as it emerges at the lower border of the pyriformis. By severing the outer end of that muscle, the latter is elevated to afford further access to the nerve as it emerges from the sciatic notch. In exposing the nerve in the thigh, the deep fascia is cut with care, as the posterior cutaneous nerve lies just below its deep surface. The biceps is displaced medially; the nerve is identified where it lies deeply and is traced distally beneath the biceps. After the nerve branches, the incision is curved downward and laterally toward the fibular head if the exposure of the peroneal nerve is desired; or the incision is curved medially and distally along the medial side of the leg if the tibial nerve is sought. Mobilization of the nerve is obtained by flexion of the knee and hyperextension of the hip. The position is maintained by a cast for several weeks until the nerve has firmly united; the cast is discarded; and the nerve is gradually stretched by straightening out the knee.

The peroneal nerve is picked up at the medial border of the biceps tendon and traced distally. The posterior attachment of the peroneus longus is severed and lifted forward, thereby exposing the branches about the fibular neck. The superficial nerve continues distally in contact with the bone in the interval between the peroneus longus and the extensor digitorum longus. Then it runs superficial to the peroneus brevis and goes obliquely forward to the anterior aspect of the ankle and the dorsum of the foot. The deep peroneal nerve proceeds distally beneath the extensor digitorum longus, then behind the tibialis anticus where it is found with and lateral to the vessels. Mobilization of the peroneal nerve is obtained by flexing the knee. If necessary, the neck of the fibula may be resected to aid in approximation.

In approaching the posterior tibial nerve, the two heads of the gastrocnemius are split apart in the mid-line, exposing the tendinous arch of the soleus beneath which the nerve and the vessels pass. The arch is cut in the mid-line, and the soleus is spread, revealing the neurovascular bundle where it lies snugly against a floor of the posterior tibial muscle. Distally, it comes to lie one fingerbreadth behind the medial malleolus, then passes beneath the medial retinaculum to enter the sole of the foot. The nerve always lies lateral to the artery. The numerous small vessels with which it is intimately associated make mobilization of the nerve difficult. First, the nerve must be isolated in the thigh, and the muscular branches stripped upward; then the knee is flexed.

OBTURATOR NERVE

Anatomy. The obturator nerve arises from the 2nd, the 3rd and the 4th lumbar ventral divisions of the lumbar plexus, which is situated retroperitoneally and behind the psoas major muscle. It descends within the fibers of that muscle and emerges from its medial bor-

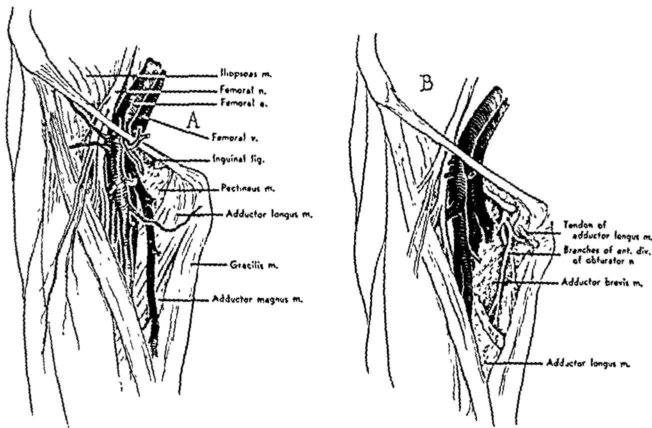


FIG. 143. Approach to the extrapelvic portion of the obturator nerve. (*Left,* superficial anatomy) The incision is made over the adductor longus tendon, which is palpable as a tight prominent cord beneath the skin. The superficial veins are ligated, and the large vessels laterally are avoided. (*Right,* intermediate level) The tendon of the adductor longus is severed and elevated, bringing into view the adductor brevis to which the branches of the anterior division of the obturator nerve intimately cling. These are resected, and the brevis is severed and elevated. The branches of the posterior division lie on the surface of the adductor magnus, which lies at the deepest level.

der near the brim of the pelvis Then it passes behind the common iliac vessels and lateral to the hypogastric vessels and the ureter; when proceeding toward its point of exit from the pelvis, it runs along the lateral pelvic wall above the obturator vessels to the upper part of the obturator foramen. While within the foramen, it divides into anterior and posterior branches.

The anterior division enters the thigh above the upper border of the obturator externus and then runs distally on the anterior aspect of the adductor brevis where it divides into branches of supply to the adductor brevis, the adductor longus and the gracilis One branch proceeds beyond as a cutaneous branch to the medial side of the thigh. Near the obturator foramen, the anterior division gives off a twig to the hip joint.

The posterior division pierces the upper part of the obturator externus and descends behind the adductor brevis and in intimate contact with the adductor magnus which it supplies. It gives off an articular branch which pierces the lower part of the magnus and supplies the back of the knee joint.

Clinical Picture. Interruption in continuity of the obturator nerve or its divisions is very rare, but when it occurs would cause paralysis of all the adductors except the magnus which has a dual nerve supply (obturator and sciatic). Disability is nil since the pectineus as well as the magnus preserves some adductor function. Sensation is reduced or absent over the inner aspect of the lower thigh. Overlap from the saphenous nerve would preserve sensation. Although the obturator nerve runs during part of its course through the psoas muscle, destruction of the nerve by infection in a tuberculous abscess is unlikely. However, irri-

tation of the nerve may cause spasm of the adductors and pain along the inner aspect of the lower thigh, signs pointing to psoas muscle involvement. This combination of spasm and pain may also occur reflexly in disease of the hip. Vague complaints of discomfort over the inner aspect of the knee should draw one's attention to the hip joint, particularly in the absence of clinical findings in the knee.

The most common involvement in the obturator nerve distribution occurs as a result of an upper motor neuron lesion, particularly in cerebral palsy. In this typical scissors gait the limbs cross over each other in marked adduction as forward progression is attempted. Passive attempts to abduct the limbs at the hips is strongly resisted, and the taut adductor longus tendon stands out prominently.

Treatment. Repair of a severed obturator nerve within the lesser pelvis is difficult and is generally unnecessary, inasmuch as some adductor function is spared, and disability is not great. When hyperactivity of the adductors interferes with gait, one or both divisions of the nerve and the adductor longus and brevis may be severed at their tendinous attachments. This procedure is described in the section on "Cerebral Palsy." Pain due to degenerative arthritis of the hip referred to the obturator nerve distribution theoretically may be relieved by section of the articular branch. However, this has not worked out too well in actual practice.

Nerve Tumors

Definitions. The ordinary benign nerve tumor is called a *neurinoma*. The constituent of the nerve from which it originates is debatable. If it arises from the connective tissue perineurium, it is designated a *perineurial fibroma* or *fibroblastoma*. If arising from the Schwann cells of the sarcolemma sheath, the proper designation is *Schwannoma*. Disregarding the site of origin, the conventional term to use is *neurofibroma*.

Types. Nerve tumors are divided into 3 types: (1) neurofibroma, (2) neurofibromatosis (von Recklinghausen's disease) and (3) neurogenic sarcoma.

Common Features of Nerve Tumors. Microscopically, these tumors display long, slender, wirelike fibers with elongated nuclei which have a tendency to be arranged in parallel rows or palisades. Palisading always suggests a nerve sheath origin. The nuclei are grouped in streams, flowing columns, or whorls. These features are lost in the more malignant tumors.

Neurofibroma. This is a round or fusiform, firm, white mass attached to the sheath of a nerve and not containing nerve fibers.

Von Recklinghausen's Disease. This consists of large numbers of tumors, especially in the fine cutaneous nerves, skin pigmentations, hypertrophy of part or all of a limb, and various bone deformities such as scoliosis. Microscopically, palisading and whorls are seen with the addition of nerve fibrils penetrating the mass.

Neurogenic Sarcoma. This arises from a nerve, probably from a pre-existent neurofibroma. Ewing believes that most fibrosarcomas are really neurogenic in origin. They are single slow-growing tumors in the intermuscular tissue. Microscopically, in addition to the palisading and whorls, curling, rippling, or waviness of the nuclei are characteristic. The malignity is demonstrated by the swelling and the pleomorphism of the cells, mitotic figures and less tendency to palisading. (See description in section on "Bone Tumors.")

POLIOMYELITIS
(Infantile Paralysis)

Poliomyelitis is an acute infectious disease caused by a virus which inflicts typical temporary or permanent destructive changes in the central nervous system and resulting in paralysis and deformities.

Etiology

The cause is a filtrable virus[20] with the following characteristics:

1. *Isolated* often from the brain and the spinal cord.

2. *Found in* nasal secretions and rectal washings of both active cases and healthy contacts Also in flies.

3 *Destroyed by* weak disinfectants. It is resistant to glycerol. It is preserved in the frozen and dried state and persists in water despite chlorination.

4. *Several strains:* Type I (Brunhilde), type II (Lansing) and type III (Leon).

[20] Ward, R: Some advances in poliomyelitic research, Am J Phys Med 31:206, 1952.

5. *Reproduced* in the Rhesus monkey.
6. *Culture* in human embryonic tissues, producing visible changes in growing cells which are specifically inhibited by immune serum.

Epidemiology

The disease is extremely spotty or epidemic and probably is spread by infected fecal matter or food. The method of transmission and portal of entry are unknown. A viremia probably is present during the incubation period before the onset of paralysis. This is suggested by the virus's being regularly found in the blood of orally infected primates, the onset of paralysis occurring from 3 to 7 days later. Usually, only 1 case occurs per family. Infants, adolescents and adults are equally affected. The disease is prevalent during the warm months. All races and classes are affected. The bulbar type commonly has a history of a recent tonsillectomy. A history of swimming and bathing is often elicited.

Relationship of Cocksackie Group of Viruses

The C viruses are frequently found in association with the poliomyelitis virus. Both are found in the human pharynx and the stools, and both are similarly distributed in nature. However, each is immunologically distinct. The C virus is isolated from paralytic and nonparalytic poliomyelitis, sewage and flies in epidemic and endemic areas and in a variety of clinical syndromes, including aseptic meningitis, encephalitis, epidemic pleurodynia, influenza, summer grippe, herpangina, etc. In poliomyelitis, the neutralizing antibody titers to poliomyelitis and C viruses rise together during convalescence. Their relationship is unexplained. C viruses are characterized by causing paralysis in newborn mice and hamsters.

Pathology[21]

The portal of entry and the method of dissemination are unknown.

The *extraneural* pathology is limited to the reticuloendothelial system, with hyperplasia and congestion of spleen and lymph nodes. The *intraneural* pathology involves chiefly the motor nerve cells. Interstitial changes consist of congestion, edema and small punctate hemorrhages.

Anterior horn neuron changes early show swelling of the cell, enlargement of the nucleus, and disappearance of the Nissl bodies. Next, the nucleus undergoes chromatolytic degeneration, and basophilic granules fill the cytoplasm. These changes may subside at this stage with reversibility and restoration of the cells. Or further degeneration, necrosis, and breaking up of the cells occurs. The interstitial tissues display marked hemorrhage, edema, perivascular mononuclear infiltration ("cuffing"), and glial invasion followed by fibrosis. The meninges also are congested and diffusely infiltrated with cells. The degenerating neurons are surrounded and absorbed by the mononuclears and replaced by glial cells.

Cellular infiltration and congestion are seen in the *posterior ganglia and the posterior nerve roots* and may be responsible for severe peripheral neuritic pains. Efferent nerve fibers from the destroyed neurons degenerate, their innervated muscles atrophy and become fibrotic. The anterior cervical and lumbar portions of the cord are affected most severely. Involvement is spotty. Each muscle is innervated from a column of cells which extend longitudinally up and down the cord. Destruction of a portion of a column leaves sufficient cells to maintain innervation to that muscle. Muscles which characteristically are supplied by shorter columns, e.g., the tibialis anterior, are more subject to permanent paralysis because of insufficient residual functioning neurons.

The *intermediate, or internuncial cell group* are regularly involved.[22] These are situated just dorsal to the anterior horn and are concerned with synaptic relays. Through these cells impulses, including those coming from higher centers, are relayed to motor neurons of the anterior horn. Control from the higher regulatory or inhibitory centers may explain the most frequent symptom of spasm which occurs in all but totally paralyzed muscles

Changes in the *higher centers* (medulla, pons, basal ganglia, tegmentum) display perivascular cuffing, lymphoid infiltration and

[21] Steindler, A.: The newer pathological and physiological concepts of anterior poliomyelitis and their clinical interpretation, J. Bone & Joint Surg 29 59, 1947.

[22] Kabat, H., and Knapp, M. E.: The use of prostigmine in the treatment of poliomyelitis, J.A.M.A 122:989, 1943.

thrombosis.[23] These changes are usually reversible and transitory. Thus, basal ganglia lesions may explain the phenomenon of in-coordination and asynergic contraction of muscles. Reversible cerebral lesions partially explain the transitory paralysis as a loss of voluntary cerebral control. The bones become slender and rarefied. Longitudinal growth is reduced in affected extremities. Fascia becomes thickened and contracted. Subluxations and dislocations occur in weight-bearing joints.

Clinical Picture

Course. Following an unknown period of incubation, generalized constitutional involvement lasting a few days, known as the "systemic phase," is manifest by fever, irritability, sweating and respiratory and/or gastro-intestinal inflammation. Next, the virus invades the central nervous system with evidence of meningeal irritation, i.e., stiff neck, stiff back, headache, vomiting, muscle tenderness and severe neuritic pains. Spinal fluid changes are now evident, and convalescent serum therapy is indicated. As the disease involves the anterior horn cells, paresis or flaccid paralysis is evident—the so-called "paralytic stage." This acute stage lasts approximately a few days to a week, the temperature subsides, and a variable amount of muscular recovery begins. This latter "convalescent stage" lasts as long as 2 years. Generally the maximum amount of improvement takes place within the first 6 months.

Systemic Stage. This stage is characterized by prodromal upper respiratory infection and/or gastro-intestinal inflammation, fever, malaise, apprehension and cervical lymphadenopathy. There are no spinal fluid changes. The patient may recover or worsen.

Stage of Meningeal Irritation (Preparalytic Stage). This stage is marked by involvement of the central nervous system, sudden onset, high fever, prostration, headache, pain in back and neck. The patient is irritable and sensitive to the touch. The child objects to being held. Very painful spasm of various muscles occurs; almost invariably the quadriceps is involved. Then course tremors, sweating and neck rigidity follow. The head-drop sign consists of backward dropping of the head as the recumbent child is lifted from the table. The back becomes rigid. Kernig and Brudzinski signs are positive. The earliest reflexes to disappear are the superficial. Next, the deep reflexes disappear in a variable and spotty manner. The *spinal fluid* exhibits a ground-glass appearance and has a cell count of about 250 per cu. mm., chiefly polymorphs early, but later lymphocytes and mononuclears. The count may vary from 10 to 1,000. Albumin and globulin are moderately increased. The sugar content is normal or increased. The fluid is sterile. It is important to note that poliomyelitis may be present without characteristic spinal fluid changes. Recovery from this stage may occur without the advent of paralysis.

Paralytic Stage. (1) *Spinal Type.* The constitutional and meningeal signs continue. To these are added flaccid muscle weakness and paralysis with reduced corresponding deep reflexes. The involvement is asymmetric and spotty. Opposing muscles may be in spasm. The extremities, the back, the abdomen and the muscles of respiration are affected.

(2) *Bulbar Type.* This type is much less frequent; often it is associated with encephalitis and runs a more fulminating course. Constitutional and meningeal symptoms are extreme. Somnolence, stupor and emesis are common. Other symptoms are nasal speech and regurgitation through the nose. Inability to swallow causes collection of mucus in the throat, and this mucus may be aspirated. The gag reflex is absent. The medullary respiratory center may be involved.

Respiratory paralysis is of two types. (1) *Spinal Type.* In this type the intercostal muscles, supplied by T 1 to 12, and the diaphragm, supplied by C 3 to 5, are paralyzed. Respiratory excursion movements are shallow, the alae nasae dilate, and the accessory muscles of respiration are used. When the diaphragm is paralyzed, "paradoxical respiration" is noted, i.e., on inspiration the abdomen sinks inward. When the intercostals are paralyzed, inspiration causes a sinking in of the chest. Fluoroscopic examination reveals the extent of diaphragm involvement. (2) *Encephalobulbar Type.* This causes arrhythmic respirations, although the intercostals and the diaphragm

[23] Peers, J. H.: Poliomyelitis induced by the Lansing strain of virus; a comparison of lesions in man and monkey, Arch. Path. 32.928, 1941.

are intact. Respiration may suddenly fail without warning.

SHORTENING OF MUSCLES. A number of muscles are increasingly irritable as demonstrated by abnormal action currents[24] If spasm persists, permanent shortening and contracture ensue If such a muscle is the antagonist to a paralyzed muscle, the latter is stretched and weakened. Typical sites of involvement of "tight" painful muscles are the back, the hamstrings and the calf.

THE ABORTIVE CASE. This is usually recognized only during an epidemic. It appears as a nonspecific illness with symptoms of a mild infectious disease with mild fever, headache, vomiting and drowsiness; the diagnosis is proved by the elevated spinal fluid protein in the convalescent stage

DIFFERENTIAL DIAGNOSIS

The following must be considered:

1. Conditions causing systemic symptoms e.g., upper respiratory infection

2. Conditions causing meningeal irritation

A. Suppurative meningitis

B. Virus encephalitis

C. Toxic encephalitis, *e.g.*, complicating pneumonia

D. Lymphocytic choriomeningitis

E. Tuberculous meningitis

F. Injury

G. Acute rheumatic fever which causes painful extremities in children

H. Trichinosis, which causes muscular pain

3. Conditions simulating paralytic poliomyelitis

A. Acute rheumatic fever—"pseudoparalysis"

B. Bone and joint inflammation

C. Scurvy

D. Radiculomyelitis of Guillain-Barré

E. Peripheral neuritis

F. Acute encephalomyelitis of virus disease; rabies vaccine

G. Botulism. Confused with bulbar poliomyelitis. However, meningeal signs are absent and spinal fluid is negative.

[24] Ransohoff, N. S.: Curare and intensive physical therapy in treatment of acute anterior poliomyelitis, Bull. New York Acad. Med (2nd Series) 23 51, 1947

TREATMENT

Preventative Treatment

VACCINES.[25, 26] Immunization will greatly diminish the incidence and, if the disease is acquired, reduce the degree of paralysis. The vaccine is prepared by culturing and propagating the 3 types separately in monkey kidney tissue, filtering, and inactivating in formalin. Then the 3 strains are pooled. Three doses of 1 cc. each are given intramuscularly with an interval of 2 to 6 weeks between the first and the second doses, and followed by a third after 7 months or longer. The vaccine should not be given during any illness, nor should it be given to members of a household where poliomyelitis has just occurred for fear of provoking paralytic complications in individuals already incubating the virus.

Convalescent serum gives temporary immunity. Instead, one may use pooled normal adult serum which has antiviral properties.

Gamma globulin is effective if given before the stage of transient viremia.

General Measures. Avoid congested areas Isolate acute cases. Eradicate insects. Emphasize cleanliness. Avoid swimming pools. Avoid fatigue

ACTIVE TREATMENT. The patient is placed at absolute bedrest in isolation, and adequate fluid intake is provided. Sedatives are contraindicated because of their depressant effect on the central nervous system. Convalescent serum is administered intravenously in adequate dosage. 60 cc. plus 1 cc. for each pound of body weight. This is repeated every 12 hours. The bed should be firm, with boards placed under the mattress. The thighs are placed in abduction, neutral rotation, slight knee flexion, and the feet at right angle to the legs. A block or a board at the foot of the bed helps to maintain this position, stimulate the standing reflex and relax the frequently involved anterior tibial muscles. The arms are placed outward to relax the deltoids and in neutral rotation. Paralyzed muscles must be

[25] Farrell, L. N., *et al* Cultivation of poliomyelitis virus in tissue culture, Canad J. Pub Health 44:273, 1953.

[26] Salk, J E Considerations in the preparation and use of poliomyelitis vaccine, J.A.M.A. 158 1244, 1955.

kept in a state of relaxation, and the resting position may be varied to meet this requirement. Heat in any form is effective for reducing muscle spasm. Spastic muscles, particularly when they are antagonists to and stretch paralyzed muscles, must be stretched often. Salicylates are the drugs of choice for relief of pain and discomfort. Narcotics are contraindicated. Avertin may be used for children, particularly as they are being introduced to the respirator.

TREATMENT OF RESPIRATORY MUSCLE PARALYSIS.[27] Paralysis of the shoulder girdle is a warning of probable respiratory muscle failure A tracheotomy is performed immediately, and the patient is placed in a respirator. The Trendelenburg position aids drainage of bronchial mucus. Respiratory negative pressure of 12 to 18 cm. water maintains adequate ventilation. Removal from the respirator should be gradual, and the patient should be trained on movements of normal respiration. The rocking or oscillating bed which alternates the Fowler and the Trendelenburg positions favors the return of normal breathing rhythm; the abdominal contents alternately push and pull on the diaphragm, producing a tidal movement of air. Accumulation of mucus in the pharynx endangers the patient, as aspiration pneumonia and atelectasis may develop. Frequent aspirations are necessary. At the first sign of dyspnea, cyanosis, rapid pulse, and rise in temperature, atelectasis is suspected and confirmed by roentgenograms. Tracheotomy, aspiration endotracheally, oxygen and the respirator are emergency measures. Ethyl alcohol may be incorporated into the oxygen circuit as an aerosol. A mucolytic agent such as Tryptar liquefies and aids expulsion of the mucus plug. Antibiotics are given.

Patients with bulbar involvement do very badly in the respirator. The irregular inefficient respirations cannot be overcome. Measures include tracheotomy, repeated aspirations, postural drainage, parenteral fluids and oxygen. The electrophrenic respirator is a device by which an intermittent electric stimulus to the exposed phrenic nerve causes rhythmic contractions of the diaphragm and inhibits the abnormal respiratory movements.

[27] Wilson, J L · Bulbar respiratory poliomyelitis, Am J. Phys Med 31·245, 1952.

PROGNOSIS

Severe widespread paralysis is often associated with high spinal fluid cell counts. Low cell counts accompany encephalobulbar disease. The mortality is high in bulbar poliomyelitis, death being due to respiratory failure. However, rapid and complete recovery is often seen in this type. Paralysis of the muscles of deglutition rarely lasts more than a month or two. Some recovery of muscle power occurs up to 3 years after the acute phase, but the maximum amount returns within the first 6 months. Complete loss of all motor neurons supplying a muscle can be strongly suspected if muscles supplied by the same and adjoining spinal cord segments are paralyzed. Thus the outlook for recovery of a completely paralyzed anterior tibial is poor if the quadriceps and the posterior tibial are likewise involved.

COMPLICATIONS

Bronchopneumonia is most common in the bulbar type. Atelectasis occurs in respiratory muscle paralysis. Contractures and deformities develop gradually. Prolonged inactivity causes mobilization of calcium, hypercalcemia, hypercalciuria, renal calculus and pyelonephritis.

CONVALESCENT CARE

An accurate chart of muscles is prepared, and their progress is noted at intervals. The following method is suggested:

0: no contraction
1: trace of contraction
2: movement without gravity
3: movement against gravity
4: movement against gravity and with slight resistance
5: movement against strong resistance
6: normal movement and strength

Muscles that are paralyzed should be kept in a relaxed position.

Absolute immobilization should be avoided if possible. The buoyancy of water aids movement of weak muscles. The Hubbard tank is useful for this. Exercises are passive at first; later they are active. Hot moist packs reduce muscle spasm and pain. Painful tight muscles should be stretched repeatedly, with the aid of curare if necessary. A paretic muscle, be-

cause it fails to develop effective tension, will tend to atrophy.[25] Therefore, inactivity retards muscular regeneration, whereas early muscle use promotes it. Therefore, early and frequent galvanic electric stimulation will encourage development of muscle size and strength. Respiratory muscles are strengthened by exercises. Coughing aids the diaphragm. Positive inflation of the lungs several times daily counteracts development of a "frozen" thorax. Tracheotomy and aspiration are continued as long as necessary. Intratracheal instillation of mucolytic agents reduces the viscosity of the mucus. Atelectasis is an ever-present danger as long as respiratory muscle weakness exists.

The Kenny Treatment

An Australian nurse, Miss Elizabeth Kenny, stated that the injurious causes of poliomyelitis are: (1) pain, (2) muscle spasm, (3) mental alienation and (4) muscle in-co-ordination. Muscles that are destined to become flaccid are those which are stretched by spasm of their antagonists. Elimination of painful spasm is accomplished by application of hot packs and proper positioning of extremities to avoid stretching. The packs cover the muscles in spasm but do not cover the large joints. A footboard to maintain the right-angle position feet preserves the standing reflex. Spasm, alienation and in-co-ordination are determined. To avoid stimulating spasm, the paralyzed muscle is moved passively without stretching the antagonist. At the earliest active motion, passive exercises are halted to permit the patient to memorize the movement, and the muscle is stimulated. Then the muscle is stretched gently to initiate the proprioceptive impulse, and the attention of the patient is drawn to the tendon insertion by lightly stroking the skin over this area. Actual re-education and exercises are not started until spasm has subsided. Fatigue must be avoided. No splints or respirators are used Massage and movement accentuate spasm and should be avoided The efficacy of this method is questionable. Because of the variable degree of spontaneous recovery of paralysis, it is impossible to draw comparisons.

Warm Salt Baths. These relieve muscle and nerve pain. The buoyancy of salt water reduces the effect of gravity and permits the few recovering muscle fibers to inaugurate contraction of the corresponding muscle.

Splints and Braces. Theoretically, these maintain the muscles in a relaxed state. A stretched muscle becomes relatively ischemic and fibrotic so that it will not respond to regenerated nerve impulses. Conversely, a splint prevents the physiologic stretch necessary to reflex contraction which maintains normal muscle tone. The present tendency is to avoid splints except where paralysis is regarded as permanent and function must be aided.

Massage is necessary to encourage circulation and is preceded by application of heat by baking, infrared or hot moist packs. Strokes are made toward the heart.

Exercises improve muscle strength. At first they are assistive, then active without and against gravity, and finally resistive. A chart of individual muscles is kept, and improvement is noted.

Rehabilitative Treatment

Principles. Deformity may be caused by an intact, spastic or contracted muscle which is the antagonist of a paralyzed muscle. For example, tight hamstrings in the presence of a paralyzed quadriceps results in a flexion deformity of the knee. The ligamentous structures tighten so that an actual contracture of the joint eventuates. Contracture of the fascial structures likewise causes deformity. The iliotibial band causes pelvic obliquity, flexion at the knee, and external rotation of the leg. Improper posture in the presence of paralyzed muscles favors contractural deformity. When the foot dorsiflexors are paralyzed, a neglected foot-drop leads to shortening of the calf muscles and contracted posterior capsule of the ankle. The most common deformities are scoliosis, knee flexion, adduction and internal rotation of the shoulder, flexion of the hips, and hyperextension of the metacarpophalangeal joints. When it becomes apparent that muscle imbalance exists, *prevention of deformity is mandatory and is accomplished* by splinting, proper positioning and stretching of antagonists. If paralysis persists, operative intervention is indicated to restore muscle power, correct deformity and

[25] Wehrmacher, W. H., Thomson, J. D., and Hines, H. M.: Effects of electrical stimulation on denervated skeletal muscle, Arch. Phys10 Med. 26 261, 1945

provide stability. Surgery can be done 6 months after the acute phase, as the maximum amount of recovery will have taken place by that time. Before the age of 10, only soft tissue surgery is permissible, because ossification is too incomplete for bone reconstruction. A poliomyelitic extremity generally has a reduced rate of growth and is atrophied. Inequality in length of the lower extremity requires temporary or permanent epiphyseal arrest. The fragile bones are susceptible to fracture, but callus formation and union are normal.

Tendon transference is done to substitute for a paralyzed muscle. The transferred muscle must have sufficient power, and its tendon should be attached to the bone as near to the insertion of the paralyzed muscle as possible. The tendon should be retained within its own sheath or that of the paralyzed muscle or should pass through the subcutaneous fat to afford a proper gliding substance. The nerves and the blood supply should be protected. All contracted tissues should be released before transference. Normal physiologic tension should be maintained.

Peabody stated that a deformity may be dynamic, i.e., caused by muscle imbalance.[29] Therefore, arthrodesis to correct this during the growth period will most likely be ineffective, because the operative forces continue, and the deformity will recur. Tendon transference is required to provide muscle balance. This may be supplemented by arthrodesis. If deformity is static, tendon transference is insufficient. A bony fusion is necessary.

Reconstructive Surgery in Poliomyelitis

Shoulder. Knowledge of the mechanism of abduction and flexion is essential to intelligent treatment. During the first 30° of abduction and 60° of flexion, the scapula finds a position of stability in relation to the humerus. The scapula shifts slightly outward to attain this position. Beyond this point the scapula moves with the humerus in a ratio of 1:2. Therefore, the loss of scapular motion decreases abduction by one third About 20° of rotation occurs at the acromioclavicular joint. Therefore, its restriction diminishes abduction. Resection of the outer end of the clavicle is valuable for increasing the range of abduction.

Abduction is performed by the deltoid while the supraspinatus holds the head firmly against the glenoid and establishes the necessary fulcrum. The infraspinous muscles pull the head downward. The scapula rotates by virtue of the force exerted by the trapezius and the serratus anterior. The clavicular portion of the pectoralis major acts with the deltoid and the supraspinatus in forward flexing of the humerus. At the same time, the serratus anterior moves the scapula forward.

The humeral head must be depressed during abduction and flexion. This is accomplished by the infraspinous muscles (subscapularis, infraspinatus, teres minor) and the long head of the biceps as its tendon passes over the humeral head.

Stability of the scapula is secured by the rhomboids and the middle portion of the trapezius.

Clinical Applications. A head depressor mechanism is essential to abduction. Transplantation of the latissimus dorsi and the teres major to the postero-inferior aspect of the greater tuberosity restores this component. Otherwise, a Nicola procedure is necessary. Only the clavicular portion of the pectoralis major, which normally functions as a flexor, can be utilized as an abductor. The levator scapulae, the rhomboids and the trapezius are necessary to scapular rotation and can be replaced by a fascial transplant from a lower cervical spinous process to the base of the scapular spine.

Restoration of motion requires adequate muscles available for transplants. Otherwise, arthrodesis is done, provided that strong scapular rotation is present. Extensive paralysis of the forearm and the hand is a contraindication to these procedures.

Serratus Anterior Paralysis. This results in displacement of the scapula medially during abduction, materially weakening that movement. Winging of the scapula is characteristic. Surgical treatment consists of transplanting the pectoralis minor tendon to the inferior angle of the scapula. The thoracodorsal nerve to the latissimus dorsi must be visualized and protected during the procedure.[30] If this muscle is not available, a fascial strip is attached at one end to the inferior

[29] Peabody, C W.: Tendon transposition; an end result study, J. Bone & Joint Surg. 20.193, 1938.

[30] Rapp, I. H.: Serratus anterior paralysis treated by transplantation of the pectoralis minor, J. Bone & Joint Surg. 36A:852, 1954.

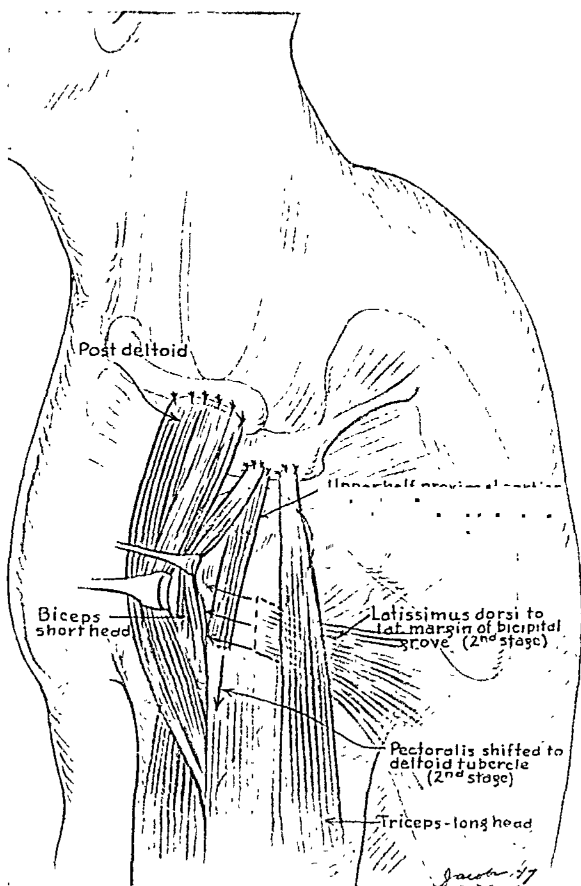


FIG. 144. Multiple muscle transplantations for deltoid paralysis. Partial paralysis of all muscles is usual; therefore, several muscles are utilized to provide strong abduction. The posterior portion of the deltoid is often spared and is transferred to a more functional position anteriorly. The operation is carried out in 2 stages:

First stage: (1) Origin of clavicular fibers of pectoralis major (upper half of muscle) transplanted to the acromion. (2) Origin of posterior deltoid shifted to acromion tip. (3) Origins of long head of triceps and short head of biceps transferred to tip of acromion.

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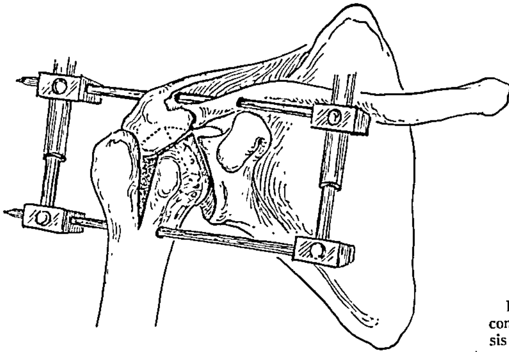


FIG. 145. Charnley compression arthrodesis of the shoulder.

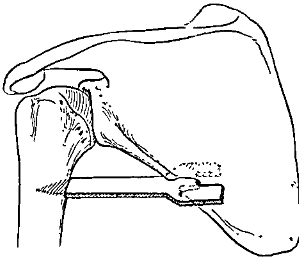


FIG. 146. Brittain compression arthrodesis of the shoulder.

scapular angle and at the other to the inferior border of the pectoralis major.

RHOMBOID AND TRAPEZIUS PARALYSIS. The scapula lacks fixation and moves forward by the unopposed action of the serratus. A fascial strip attached at the inferior scapular angle is attached to the thoracic spine or the spinal muscles medially. The insertion of the levator scapulae at the superior angle is transferred to a more forward position to elevate the acromion, thereby restoring the lost function of the upper trapezius.

DELTOID PARALYSIS. With the arm elevated, fascial strips connect the trapezius with the deltoid insertion. First, the humeral head must be stabilized by attaching the tendons of the latissimus and the teres major to the postero-inferior aspect of the greater tuberosity. An alternative is to perform the Nicola procedure. If the posterior portion of the deltoid remains active, it may be detached and transferred to a more favorable position at the outer end of the acromion.

MULTIPLE MUSCLE TRANSPLANTATIONS.[31,32] The usual presenting situation is a varying

[31] Harmon, P. H: Surgical reconstruction of the paralyzed shoulder by multiple muscle transplantations, J. Bone & Joint Surg. 32A:583, 1950.

[32] Ober, F. R: Transplantation to improve function of the shoulder joint, Instructional Course Lectures, Am. Acad. Orth Surg, Ann Arbor, Mich., Edwards, 1944.

FIG. 144. (*Continued from facing page*) Second stage: about 3 weeks later. (1) Insertion of clavicular fibers of pectoralis major transferred to region of deltoid tubercle. (2) Insertions of latissimus dorsi and teres major transplanted over lateral surface of the humerus to the lateral margin of the bicipital groove.

(Harmon, P. H.: Surgical reconstruction of the paralytic shoulder by multiple muscle transplantations, J. Bone & Joint Surg. 32A:583)

degree of involvement of many muscles about the shoulder. Therefore, transplanting only one muscle to restore deltoid function is doomed to failure. It becomes necessary to utilize all available muscles. A prerequisite is effective scapular control by a functioning trapezius and serratus anterior, or an effective muscle or fascial substitute. Often the posterior portion of the deltoid is preserved and is transferred to a more forward position. In addition, the clavicular portion of the pectoralis major, the short head of the biceps and the long head of the triceps are transferred to the acromion. At the second stage, the insertion of the clavicular head of the pectoralis major is transferred to the deltoid tubercle to function more efficiently as an abductor. At the same time the latissimus dorsi insertion into the medial side of the bicipital groove is removed, and the tendon is rerouted about the humerus and attached to the lateral margin of the groove. This furnishes the external rotator.

Arthrodesis of the Scapulohumeral Joint. The humerus should be fixed in no more than 45° of abduction in the adult and a little higher in the child. In addition, about 30° of forward flexion and sufficient external rotation so as to place the forearm about 30° above the horizontal provides the most useful position. Good power of scapular rotation is a

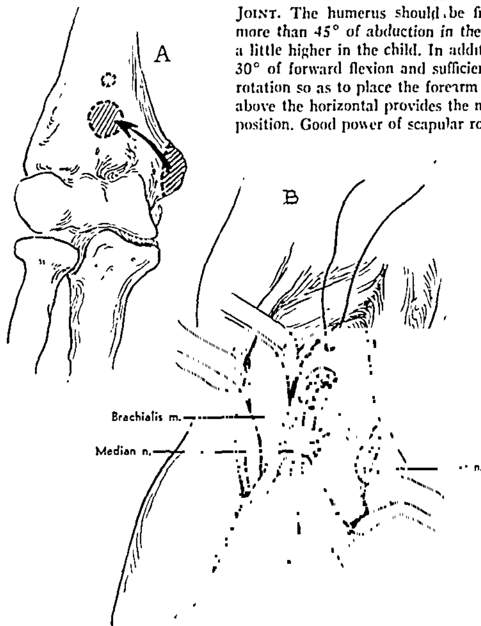


Fig. 147. The Steindler operation to restore active flexion to the elbow The common tendon origin of the flexor-pronator group is removed with the medial epicondyle and transferred to a higher level.

necessary prerequisite. If the serratus is paralyzed, the trapezius alone can abduct the shoulder to 45°. The range of motion is generally greater if arthrodesis is done before the age of 12. Epiphyseal growth is not endangered.

Elbow. When the brachialis anticus, the biceps and the brachioradialis muscles are weakened or paralyzed, active flexion must be reinforced or completely restored. The Steindler operation consists of removing the common tendon of origin of the pronator radii teres, the flexor carpi radialis, the palmaris longus and the flexor carpi ulnaris and transferring it to a higher point on the humerus.[33] The method is rendered more effective by inserting a bone block posteriorly at the lower end of the humerus, thereby limiting extension of the elbow to 90°. Active extension of the elbow is necessary only when crutches must be used and when pushing motions of the extremity are desirable. The brachioradialis origin may be transposed to the triceps or the humerus posteriorly.[34]

[33] Steindler, A.: Muscle and tendon transplantation at the elbow *in* Reconstruction Surgery of the Extremities, Am. Acad. Orth. Surg., Instr. Course Lectures, Ann Arbor, Mich., Edwards, 1944

[34] Ober, F. R., and Barr, J. S.: Brachioradialis muscle transfer for triceps weakness, Surg., Gynec. & Obst. 67:105, 1938.

Forearm. Fixed pronation is the main disabling deformity. *Supinator function may be* restored by tendon transference to the dorsal and the radial aspects of the radius. The pronator teres and the flexor carpi radialis are commonly used. Their tendons are sutured together and routed subcutaneously about the ulnar side of the forearm, then dorsally across to the opposite side, and fixed to the radius on the radial and the volar aspects.[35]

Wrist and Hand. As a rule, following poliomyelitis both flexors and extensors are involved, and rarely is the paralysis so isolated that a sufficiently strong muscle is available for transference. Arthrodesis of the wrist will overcome a flexion deformity and provide a mechanical advantage for weakly functioning finger flexors. If wrist extensors are functioning, these may be utilized for transfer to the finger flexors. Restoration of opposition to the thumb is necessary for pinch and grasp. The subject is discussed more fully in the section on "The Hand."

Hip. Gluteus Medius Paralysis. The gluteus medius, acting as an abductor, is the most important hip stabilizer, elevating the opposite end of the pelvis when weight is borne

[35] Tubby, A. H.: Deformities and Diseases of Bones and Joints, vol. 2, New York, Macmillan, 1912.

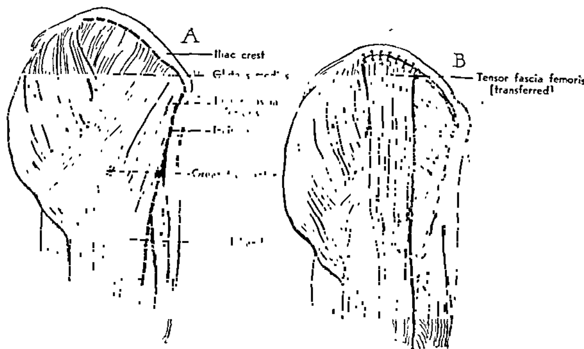


Fig. 148. Legg procedure for correcting gluteus medius weakness. The origin of the tensor fasciae femoris is transferred posteriorly. (Redrawn from Legg, A. T.: New England J. Med. 209:61)

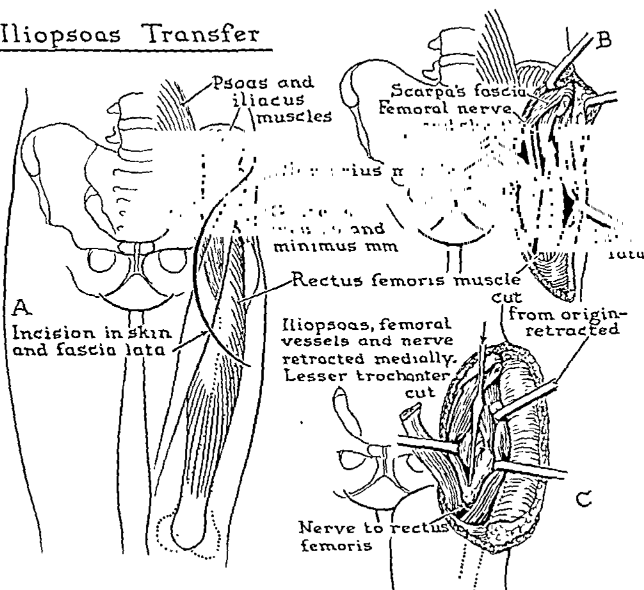
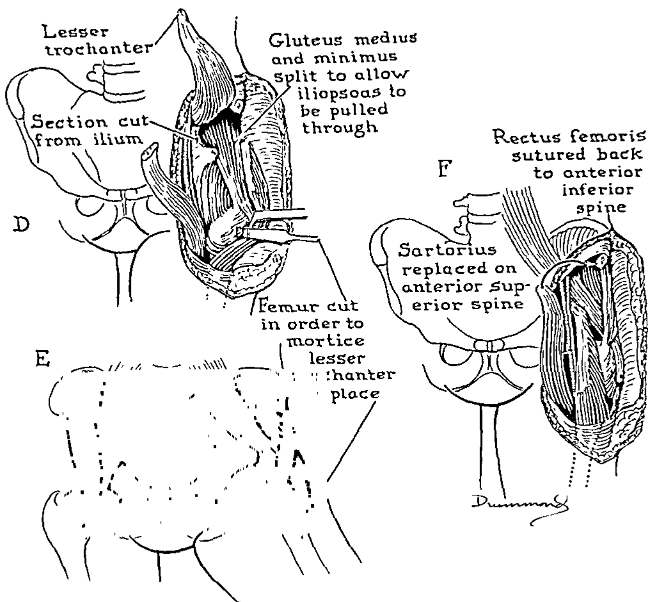


FIG. 149. (Mustard, W. T., Iliopsoas transfer for weakness of hip abductors; preliminary report, J. Bone & Joint Surg. 34A:647-650)

on the corresponding extremity. When this muscle is paralyzed, the pelvis loses its support and sags, the trunk listing toward the affected side in order to attain balance. The result is a typical side-lurching gait. Transference of functioning muscle to the greater trochanter restores abductor power and eliminates the limp. Available muscles are the iliopsoas, the erector spinae and the tensor fascia femoris Only the iliopsoas possesses sufficient strength to replace a paralyzed gluteus medius. The others are useful to reinforce a partially weakened muscle. A slightly weakened gluteus medius can be improved by transferring the origin to a point more posteriorly on the iliac crest.

ILIOPSOAS TRANSFER.[36] Prerequistes are (1) a normal sartorius to act as a hip flexor, (2) a good or normal gluteus maximus to assist in pelvic stability, and (3) good lateral abdominal muscles which maintain lateral stability of the pelvis. The hip is approached by a Smith-Petersen incision The origins of the sartorius and the rectus femoris are removed. The femoral nerve and vessels are retracted medially. The insertion of the iliopsoas at the lesser trochanter is severed, removing a flake of bone with the tendon. Tendon and muscle are reflected upward. A notch is cut in the ilium

[36] Mustard, W. T: Iliopsoas transfer for weakness of the hip abductors, J. Bone & Joint Surg. 34A:647, 1952.



between the superior and the inferior iliac spines large enough to accommodate the muscle belly, which is drawn through and attached to the greater trochanter. Postoperatively, the hip is maintained in abduction for several weeks until the new attachment is secure.

GLUTEUS MAXIMUS PARALYSIS. This, too, provides stability of the hip. Its loss is manifested by the body's lurching backward with weight-bearing. The erector spinae and the tensor fascia femoris are used as transfers.

ERECTOR SPINAE AND TENSOR FASCIA FEMORIS TRANSFER.[37] Through a longitudinal incision in the lumbar area alongside the spinous processes, the erector spinae is exposed. It is split in two, and the outer half is freed from its attachments to the sacrum and the iliac crest and mobilized upward. Another incision is made longitudinally along the lateral aspect of the thigh, and a strip of thickened fascia lata (iliotibial band) is cut and dissected upward into a portion of the tensor fascia femoris. Then the flap is carried posteriorly beneath the remaining tensor fascia femoris and below the greater trochanter, then upward beneath the fascia of the gluteus maximus to emerge at the upper wound. There it is attached under tension to the free end of the erector spinae. Postoperatively, the hip must be maintained for several weeks in full extension.

[37] Ober, F. R.: An operation for relief of paralysis of the gluteus maximus muscle, J.A.M.A. 88.1063, 1927.

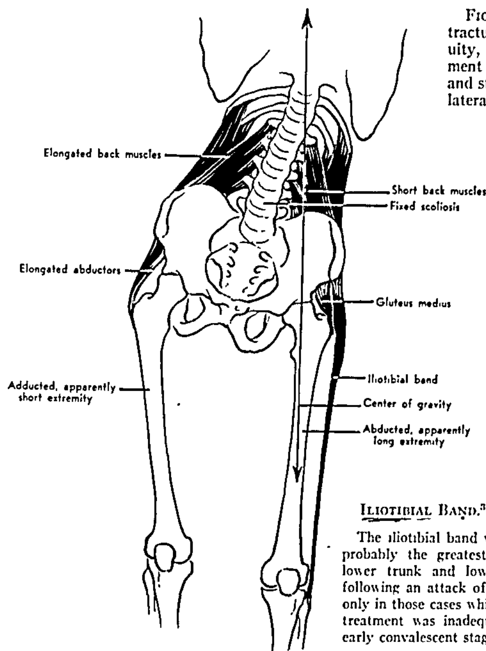


FIG. 150. Iliotibial band contracture, showing pelvic obliquity, lumbar scoliosis, displacement of the center of gravity, and stretched, elongated contralateral muscles.

FLAIL HIP. Frequently, this is associated with subluxation or complete dislocation. Although the dislocation is easily reduced, it recurs because the acetabulum is shallow. Arthrodesis provides complete stability. The prerequisites for fusion are (1) adequate abdominals and quadratus lumborum, (2) sound knee ligaments, (3) stable foot and ankle and (4) a functioning opposite lower extremity In the case of bilateral dislocation, shelf stabilization is preferred.

DISLOCATED HIP WITH RESIDUAL MUSCLE POWER. Reconstruction of the hip by deepening the acetabulum and formation of a shelf should be followed by muscle transfer to restore abductor power.

ILIOTIBIAL BAND.[38] Irwin states:

> The iliotibial band with its allied structures is probably the greatest deforming factor in the lower trunk and lower extremity involvement following an attack of poliomyelitis This is true only in those cases which had no care or in which treatment was inadequate during the acute and early convalescent stage[39]

Pathologic Anatomy. The iliotibial band is a thickened portion of the fascia lata along its lateral aspect. The fascia lata arises from the coccyx, the sacrum, the iliac crest, Poupart's ligament and the pubic ramus. Between two layers it encloses the gluteus maximus and the tensor fascia femoris, giving attachment to the latter muscle and most of the former. The fibers of the fascia converge to form the iliotibial band along the lateral side of the thigh. It is continuous medially with the lateral intermuscular septum which attaches to the linea aspera. Distally, it gives origin to the

[38] Yount, C. C: The role of the tensor fascia femoris in certain deformities of the lower extremity, J Bone & Joint Surg 8·171, 1926

[39] Irwin, C. E Orthopedic Correspondence Club Letter, March 6, 1947.

short head of the biceps. At the level of the knee joint, it spreads out and attaches to the lateral tibial condyle and the head of the fibula. The iliotibial band lies in a plane anterior to the hip joint and posterior to the knee.

Involvement of the attached muscles is responsible for the increased tension under which it is placed during the acute and the convalescent stages. The taut band is perceived by deep palpation while adducting and extending the thigh. Spasm in the gluteus maximus is demonstrated by resistance to passively flexing the hip while the knee is fully extended. Spasm in the short head of the biceps is demonstrated by resistance to extension of the knee while the hip is flexed. The patient assumes the most comfortable position in which the thigh is flexed, abducted and externally rotated at the hip while the knee is flexed. This relaxes tension on the band. If tension is not overcome by stretching during the acute stage, band contracture becomes progressive, and permanent deformity ensues:

1 *Flexion and abduction contracture of the hip,* because the band lies in a plane anterior and lateral to the hip.

2. *External rotation of the thigh.* The external rotators of the hip become contracted in the position which the patient assumes for comfort

3. *Genu valgum and flexion contracture of the knee.* The compression forces acting upon the outer half of the epiphyseal plates probably retard longitudinal growth laterally while growth on the medial side continues unimpeded. The band lying in a plane posterior to the knee exerts a flexing force.

4. *External rotation of the tibia.* The direction of the lowermost fibers obliquely downward and forward to insert on the anterolateral aspect of the tibia produces a torsional force. The tibia may subluxate posteriorly.

5. *Short leg.* This may be due to compression forces acting upon the lower femoral and the upper tibial epiphyseal plates.

6 *Varus deformity of the foot.* Because the axes of the knee and the ankle do not lie in the same horizontal plane, any attempt to fit an ordinary brace to such an extremity will force the foot into varus.

7. *Pelvic obliquity, increased lumbar lordosis, scoliosis.* When the extremity is forced into the weight-bearing position, the contracted hip will incline the pelvis forward and throw the lumbar spine into increased lordosis. The opposite end of the pelvis will displace upward, and the lumbar spine deviates toward the affected hip.

Treatment. Prevention demands early energetic stretching of the iliotibial band during the acute and the convalescent stages. Treatment is continued until the hip and the knee joints can be carried through their full range of motion. The full length of the band is thereby preserved.

Surgical treatment is necessary when increased tension becomes a progressive contracture. A section of the iliotibial band and its attached intermuscular septum must be resected and the deformities corrected by traction and casts.

Technic. Two short leg casts from the tibial tubercle to the toes are applied the day before operation. At surgery a portion of the tight band at a point 3 or 4 inches above the knee is removed, and a portion of the fascia lata covering the vastus lateralis is divided transversely. The lateral septum is divided. A Kirschner wire is passed transversely through the supracondylar area of the opposite femur, and the short leg casts are extended to the upper thigh incorporating the wire. The affected thigh is held in abduction and flexion, and traction is applied to the Kirschner wire to bring the pelvis to the horizontal level. The body portion of a spica cast is applied and fixed to the well-leg cast. The trunk, the pelvis and the well leg are now held securely. Next, the contracted leg is adducted, extended and internally rotated until considerable resistance is felt, and the double spica is completed. Further wedging of the cast on the operated side at intervals of 3 to 5 days secures complete correction.

A Soutter fasciotomy will not correct the hip contracture, because the rectus femoris and the iliopsoas do not contract until late in flexion deformities at the hip; the gluteus medius and minimus contract late in abduction contracture. It is only in very late neglected contractures that a Soutter procedure should be done in addition to resection of the iliotibial band.

FIXED PELVIC OBLIQUITY. When iliotibial band contracture and the consequent tilted

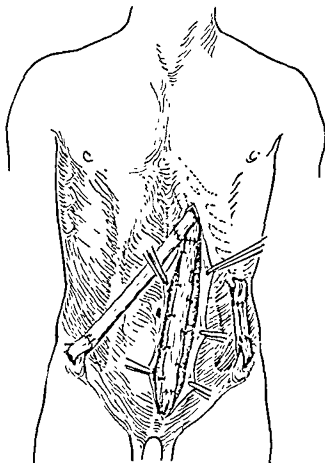


FIG. 151. Fascial transplants for paralyzed abdominal muscles. One transplant from the lower costal margin to the opposite anterior superior iliac spine, another from the lower costal margin to the symphysis, and the third from the lowermost rib laterally to the middle of the iliac crest represent the different types used. Various combinations are employed, depending on the site of abdominal muscle deficiency The lateral abdominal fascial transplant is useful for overcoming quadratus lumborum paralysis associated with paralytic scoliosis and pelvic obliquity.

pelvis and ipsolateral scoliosis have lasted a long time, adaptive bony changes, particularly of the spine, render the deformity permanent and not amenable to release of contracted soft tissues. The affected limb is abducted and apparently long The opposite hip is adducted and apparently short; its hip abductor is stretched and weakened. The lumbar spine deviates toward the contracted side so that the trunk and therefore the center of gravity comes to lie over and beyond the affected hip. The contralateral trunk muscles become elongated, and their contractility is impaired. The patient must widely abduct the affected extremity in order to take a step. Obviously, the adducted position, weakened abductors and stretched lateral trunk muscles on the opposite side make it difficult to attain balance and to bring the "long" extremity forward when walking. (This is similar to the disability caused by severe gluteus medius paralysis.) To overcome this situation it is necessary to obtain abduction of the "short" extremity, regain apparent length and bring the weight-bearing line nearer the center of the body. This is accomplished by subtrochanteric osteotomy and abduction of the distal segment.

Scoliosis per se is a lesser cause of pelvic obliquity. Rarely, weakness of the lateral abdominals may be a causative factor.

Treatment. Roentgenologic studies of the lumbar spine determine whether the pelvic obliquity and scoliosis are fixed. The subtrochanteric region of the femur is exposed. A heavy Steinmann pin is inserted just below the greater trochanter. Below this a wedge-shaped section of bone is removed with the base directed laterally, the medial cortex of the femur being left intact to avoid displacement. The removed bone is broken up into small fragments and replaced in the defect. A double spica cast is applied, incorporating the pin. After 4 weeks a wedge is removed from the cast below the level of the pin, and the distal limb is abducted as the medial cortex is fractured. The soft callus about the osteotomy site prevents displacement. The cast is repaired with plaster, and immobilization is continued for 8 more weeks

When pelvic tilt is extreme and the trunk is shifted markedly over the contracted hip, subtrochanteric osteotomy and adduction of the femur are indicated.

ABDOMINAL MUSCLE PARALYSIS. Insufficiency of the anterior abdominal muscles results in abdominal bulging, increased lumbar lordosis, and inadequate bowel and bladder expulsion. Stability of the pelvis in front necessary to flexion of the hips is lessened. When the rectus abdominis is paralyzed, the pelvis tilts anteriorly with consequent increase of lumbar lordosis. Rectus weakness is identified by Beevor's sign: when the head is raised against resistance, the umbilicus shifts toward the contracting muscles. If both recti are inactive, the umbilicus remains immobile. Weak-

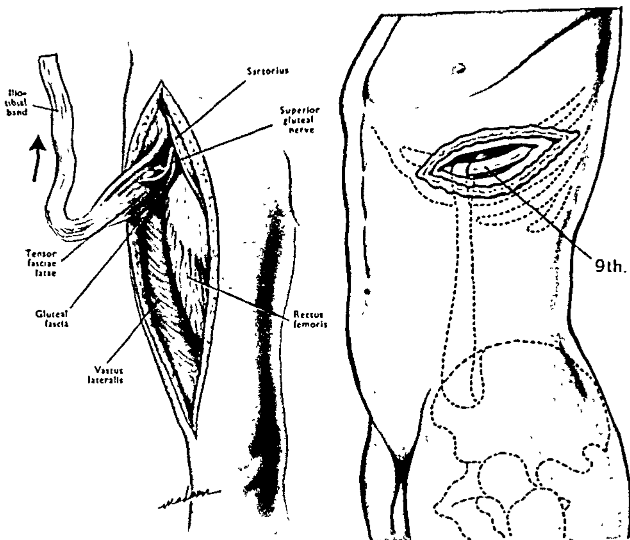


FIG. 152. (*Left*) Isolation and mobilization of musculotendinous transplant.
FIG. 153. (*Right*) Transposition of musculotendinous transplant.
(Clark, J. M. P., and Axer, A.: Muscle-tendon transposition for paralysis of abdominal muscles, J. Bone & Joint Surg. 38B:475-484)

ness of the transversus and the obliques is diagnosed by inability to initiate the movement of turning over. Paralysis of the quadratus lumborum results in loss of stabilizing of the lateral pelvis while abducting the hip The pelvis tilts downward, and pelvic obliquity is the consequence. The lumbar spine deviates toward the ipsolateral side. Quadratus lumborum weakness is identified by inability to pull the pelvis proximally on one side against resistance offered by the examiner pulling the extremity distally.

FASCIAL TRANSPLANTATION.[40, 41] Strengthening the abdominal wall and stabilizing the spine and the pelvis against the deforming influence of unopposed functioning muscles is possible only by transplanting strips of fascia. Fascia lata reinforces or entirely replaces the insufficient muscle, and not only hypertrophies but grows in length. After several years it is visible as a prominent subcutaneous band. The following procedures are employed most commonly:

Rectus and Oblique Weakness. Two strips of fascia, each one measuring 1 by 9 inches, are placed subcutaneously in crisscross fashion across the abdomen. Each is fastened at its proximal end to the periosteum and the soft tissues about the lower costal margin. The distal end is fastened to an osteoperiosteal tunnel in the opposite iliac crest

[40] Lowman, C L · Plastic repair for paralysis of abdominal musculature, New England J. Med. 205. 1187, 1931.

[41] Mayer, L.. The significance of the iliocostal fascial graft in the treatment of paralytic deformities of the trunk, J Bone & Joint Surg 26 257, 1944.

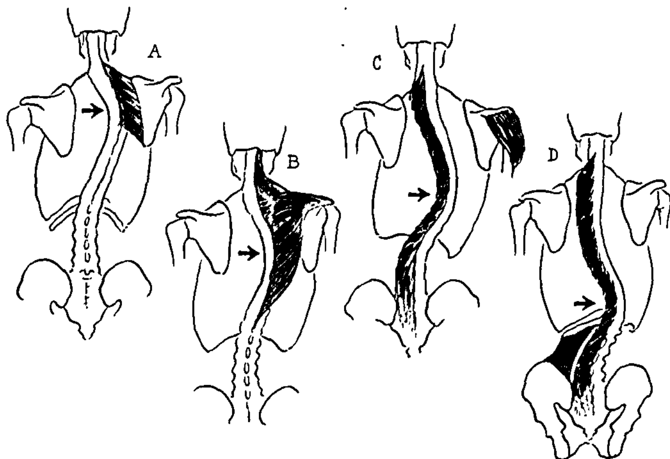


FIG. 154. Paralytic scoliosis, demonstrating the effect of unequal muscle pull. (A) Rhomboid muscle paralysis. (B) Trapezius muscle paralysis. (C) Sacrospinalis and deltoid paralysis. (D) Sacrospinalis and quadratus lumborum paralysis. (Redrawn from Kleinberg, S.: Scoliosis, Baltimore, Williams & Wilkins)

near the anterior superior spine. When only the lower portion of a rectus is paralyzed, a fascial strip is extended from the functioning muscle above to the pubic symphysis below If the upper rectus is deficient, the strip extends from the functioning muscle below to the xiphoid above. Paralysis of the obliques is demonstrable by localized abdominal bulging. The fascial strip is fixed to the rectus aponeurosis near the umbilicus The other end is attached to the costal margin above or the iliac crest below as the case may be.

Lateral Abdominal Wall Paralysis and Pelvic Obliquity A wide band of fascia lata is attached to the 9th rib above and to the anterior iliac crest and Poupart's ligament below; or the tensor fascia lata, with iliotibial band attached may be transposed upward to the 9th rib.[42]

Quadratus Lumborum Paralysis. The fascial strip is attached above to the erector spinae at the level of the dorsolumbar junction and below to the posterior half of the iliac crest.

PARALYTIC SCOLIOSIS.[43] Postpoliomyelitic scoliosis is due to asymmetric paralysis of paraspinal muscles. The imbalance is a continually operating mechanism, thereby explaining the development of scoliosis in spite of continuous recumbency. However, the upright position aggravates the curve by adding the factor of gravity. When paralysis is extreme and symmetric, scoliosis may not develop. Spinal curvatures may be divided into 2 types:

1. *Convexity of curve toward stronger muscle groups.* Examples are the iliopsoas, the gluteus medius, the gluteus maximus, the latissimus dorsi, the rhomboids and the deltoid.

2. *Concavity of curve toward stronger muscle groups.* Examples are the abdominals, the sacrospinalis, and the quadratus lumborum.

[42] Clark, J. M P, and Axer, A Muscle-tendon transposition for paralysis of abdominal muscles, J. Bone & Joint Surg. 38B.475, 1956

[43] Kleinberg, S.: Scoliosis, Baltimore, Williams & Wilkins, 1951.

Contracture of the pelvitrochanteric muscles and the iliotibial band with resultant fixed pelvic obliquity deviates the spine toward that side.

A theoretic added factor is the neurogenic element which results in structural changes of the spine. Five per cent of poliomyelitis patients develop scoliosis.

The curvature, contrary to the idiopathic type, may develop and progress even after growth is completed. It may make its appearance at any time within 10 years after the acute phase and tends to become severe. Therefore, the postpoliomyelitic patient should be checked every 6 months for several years. Three major types of curve are seen: high cervicodorsal kyphoscoliosis, the long dorsolumbar scoliosis and the lumbar curve.

Treatment. Preventive treatment consists of prolonged recumbency, as long as 6 months, in patients with paralysis of trunk and abdominal muscles. The spine should be x-rayed in the standing position every 3 months. Recumbency is maintained until stability of the curve is attained, or progression indicates the need for correction. Lying on a concave frame favors the weak abdominals. Upon resuming ambulation, if asymmetric involvement of the abdominals and the hip muscles exists, the use of crutches with a tripod gait is mandatory. An alternating gait is permissible only if one is certain that the asymmetrically involved trunk muscles will not increase the scoliosis.

Surgical treatment consists of correction of the curve, and fusion and is particularly indicated in young patients with gross asymmetric involvement and a rapidly progressing curve. Early fusion should be avoided, as a lordosis may result. Many paralytic curves become static and stable and require no fusion. When the curve is recent, and isolated muscle involvement can be detected, special methods are available to halt further progression of the curve:

PELVIC OBLIQUITY is treated by iliotibial band resection (q.v.)

ABDOMINALS AND THE QUADRATUS LUMBORUM are treated by fascial transplants (q.v.)

SCAPULAR ELEVATOR MUSCLES. For high cervicothoracic curves, two strips of fascia are attached to the scapular spine. One is fixed to the cervical muscles at the apex of the curve on the concave side; the other is attached to the spinous process of the first thoracic vertebra.

RHOMBOIDS AND THE LEVATOR SCAPULAE normally pull the scapula upward and inward and exert tension on the upper 4 dorsal and cervical vertebrae. When these are paralyzed, the spine is pulled to the opposite side. Fascial transplants are attached to the vertebral border of the scapula and into the spinal muscles and the latissimus dorsi.

Knee. When the knee is fully extended, it is stable even in the absence of quadriceps function. The position is favored while standing by equinus of the foot. Loss of active extension of the knee in the presence of strong hamstring flexors favors development of a flexion contracture. The opposite situation of a strong extensor and weak flexors encourages a recurvatum deformity—in reality hyperextension. A strong biceps (which attaches to the fibular head) plus weakened other hamstrings and quadriceps results in flexion, abduction (valgus), and external rotation of the leg on the thigh. The same deformity is produced by a contracted iliotibial band which attaches to the head of the fibula and the lateral tuberosity of the tibia. Persistence of deformity during the growth period encourages the bony structures to develop in adaptation, and the deformity is rendered permanent.

Quadriceps paralysis is the only indication for tendon transference about the knee. All deformity must first be corrected. Slight flexion contracture can be overcome by wedging casts. Severe flexion deformity is treated by supracondylar femoral osteotomy. Subcondylar osteotomy of the tibia will correct a genu valgum and tibial torsion. Flexion contracture of the hip and Achilles tendon contracture must first be overcome. The biceps—the strongest hamstring—is utilized most frequently for transfer to the patella. Occasionally, the semitendinosus is added. The other hamstrings are too weak to be effective. It is necessary that some flexion power be preserved by at least one functioning hamstring and gastrocnemius; and the hip muscles must be functioning

TRANSFER FOR QUADRICEPS PARALYSIS. Through a lateral longitudinal incision the biceps muscle and tendon are exposed to its insertion to the head of the fibula. The common peroneal nerve which skirts the medial border of the biceps tendon is isolated and re-

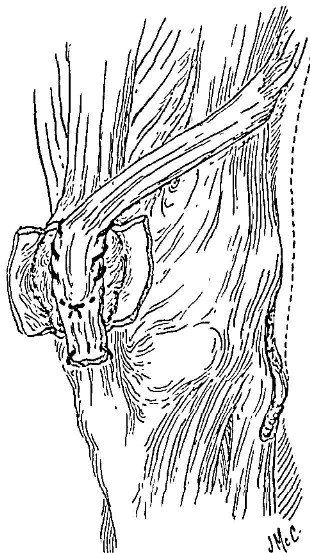


FIG. 155. Transference of biceps femoris for paralysis of the quadriceps. The semitendinosus may be similarly transferred from the medial side to reinforce extensor power.

tracted. The tendinous insertion is severed, and tendon and muscle are freed up to the point of entrance of nerves and vessels into the muscle belly. The tendon is routed subcutaneously obliquely forward The iliotibial band and the intermuscular septum are resected to provide free access of the muscle to the anterior thigh. Through a medial longitudinal incision, the tendon of the semitendinosus is likewise severed at its insertion, freed upward and routed subcutaneously forward Both tendons are embedded within a tunnel gouged from the anterior aspect of the patella and sutured to the soft tissues. Postoperatively, a spica cast is applied with the hip fully extended to relax the hamstrings At 3 weeks, exercises may begin. A brace is worn to maintain extension until the transferred muscles acquire adequate power.

GENU RECURVATUM. The mechanisms by which backward bowing of the knee is produced in poliomyelitis are:

1. *Quadriceps paralysis,* regardless of normal hamstrings and calf muscles. Constant attempts at locking the knee in full extension lead to backward bowing of the upper end of the tibia. Treatment consists of osteotomy of the upper tibia and transfer of a hamstring to the patella.

2. *Hamstring and triceps surae paralysis,* which leads to stretching of the posterior capsule. A portion of the biceps tendon laterally and of the gracilis and the semitendinosus tendons medially may be transplanted to a posterior position on the femoral condyles to act as restraining ligaments. However, these soft tissues eventually stretch and deformity recurs. The only alternatives are arthrodesis or a brace.

COMPLETE PARALYSIS ABOUT THE KNEE. This results in a flail joint. Wearing a long leg brace with locked knee joint or arthrodesis overcomes the disability.

Foot. The correction of deformity can be accomplished by reconstruction and arthrodesis. If the deformity is due to muscle imbalance, arthrodesis in a growing child will not prevent a recurrence. Restoration of muscle balance is necessary. The requisites for satisfactory transfer are: (1) the transferred muscles or muscles must be *equal in power* to the paralyzed muscle; (2) the tendon must pass in a *direct line* from the muscle to the point of insertion, (3) passages must be surrounded by a tissue which will not adhere and restrict *gliding,* preferably a tendon sheath or subcutaneous fat; (4) *pulleys* are fully utilized for maximum mechanical efficiency, e.g., the transverse crural ligament; (5) the muscle must be under *normal physiologic tension;* (6) *nerve and blood supply* must be protected, (7) it must be attached to the tendon or actual *point of insertion* of the paralyzed muscle, or to the point which will restore balance and overcome deformity; and (8) *contracted structures must be overcome,* and necessary *bone operations must precede* tendon transfer.

TENDON TRANSFER ABOUT THE FOOT. This is required for loss of power of the evertors



Fig. 156. Calcaneus foot. (*Left*) Dorsiflexion apparently exaggerated. (*Right*) Abnormally high longitudinal arch.

(peronei), invertor (posterior tibial), dorsiflexors (anterior tibial, extensor hallucis longus) and plantar flexors (gastrocnemius, soleus). A variety of combinations of paralyses occur, and each case must be judged individually.

1. *Extensor-invertor insufficiency* leads to to equinus and planovalgus deformity. The extensor hallucis longus is transposed to the base of the first metatarsal, and the interphalangeal joint of the great toe is arthrodesed. Or the peroneus longus tendon is transferred to the dorsum of the foot at the first cuneiform.

2. *Evertor insufficiency* causes varus deformity and elevation of the base of the first metatarsal, accentuating the longitudinal arch. The extensor hallucis longus or tibialis anticus is transferred to the outer border of the foot at the base of the 5th metatarsal bone. After the age of 10, evertor or invertor weakness requires in addition a stabilization operation.

3. *Calf Muscle Insufficiency.* Progressive calcaneus deformity results. The tibialis anticus and the peronei are transposed to the

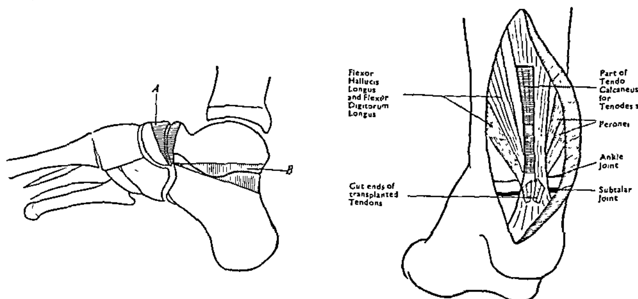


Fig. 157. Elmslie's operation: (*Left*) Diagram showing wedges of bone removed: (A) at first stage; (B) at second stage. (*Right*) Diagram showing transplanted tendons. (Cholmeley, J. A.: Elmslie's operation for calcaneus foot, J. Bone & Joint Surg. 35B:46)

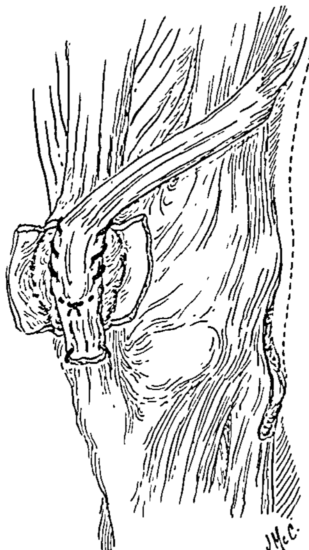


FIG. 155. Transference of biceps femoris for paralysis of the quadriceps. The semitendinosus may be similarly transferred from the medial side to reinforce extensor power.

tracted. The tendinous insertion is severed, and tendon and muscle are freed up to the point of entrance of nerves and vessels into the muscle belly. The tendon is routed subcutaneously obliquely forward. The iliotibial band and the intermuscular septum are resected to provide free access of the muscle to the anterior thigh. Through a medial longitudinal incision, the tendon of the semitendinosus is likewise severed at its insertion, freed upward and routed subcutaneously forward. Both tendons are embedded within a tunnel gouged from the anterior aspect of the patella and sutured to the soft tissues. Postoperatively, a spica cast is applied with the hip fully extended to relax the hamstrings At 3 weeks, exercises may begin. A brace is worn to maintain extension until the transferred muscles acquire adequate power.

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1. *Quadriceps paralysis,* regardless of normal hamstrings and calf muscles. Constant attempts at locking the knee in full extension lead to backward bowing of the upper end of the tibia. Treatment consists of osteotomy of the upper tibia and transfer of a hamstring to the patella.

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TENDON TRANSFER ABOUT THE FOOT. This is required for loss of power of the evertors

ASTRAGALECTOMY. Complete excision of the astragalus is useful for severe calcaneal deformity and flail feet in children before the age at which foot arthrodesis can be performed. The deformity is corrected, and the distal end of the tibia articulates directly with the calcaneus and the scaphoid. The range of dorsiflexion and plantar flexion is small, and lateral motion is nil, so that good stability is afforded. However, the foot is weakened, the extremity shortened, and a painful pseudarthrosis may ensue. If the triceps surae or the tibials are functioning, the procedure is contraindicated, as equinus or varus deformity, respectively, will result. If adequate functioning muscle is available, tendon transference is more desirable as a temporary measure until stabilization can be done at a later date.

LOSS OF DORSIFLEXOR POWER AT THE ANKLE (DROP FOOT; PARALYTIC EQUINUS). Paralysis of the anterior group of leg muscles results in inability to dorsiflex the foot actively. The Achilles tendon secondarily becomes contracted. The equinus must be overcome for proper ambulation. When calf muscles as well as the anterior group are paralyzed, a flail ankle results, and fusion of the ankle is indicated (see Panastragalar Arthrodesis). If calf muscles are sufficiently strong to aid propulsion, the equinus can be prevented by (1) a posterior bone block operation, or (2) a tarsal bone plastic procedure of the Lambrinudi type. A triple arthrodesis and Achilles tendon lengthening are also done, although the latter procedure may be condemned on the ground that the calf muscle may be weakened. Complete correction of the equinus is not desirable when quadriceps paralysis is present.

POSTERIOR BONE BLOCK OF CAMPBELL.[44] A longitudinal incision along the medial border of the Achilles tendon extends upward from the os calcis for several inches. The tendon is lengthened, if necessary, by a Z-plasty. By blunt dissection the posterior surface of the lower tibia is exposed without opening the periosteum. The flexor hallucis tendon is retracted medially. Upon passively dorsiflexing the foot, the posterior capsule of the ankle and the subastragalar joints are rendered taut. These are cut transversely. The posterior edge of the astragalus is visible and is resected. Below this a wedge-shaped cavity is gouged from the os calcis into which a strong cortical graft is inserted so that it extends upward and abuts against the posterior surface of the tibia. This is reinforced by surrounding it with multiple small pieces of cancellous bone. The soft tissues are closed snugly about the grafts, and a cast is applied, the ankle being positioned at a right angle.

TARSAL PLASTIC OF LAMBRINUDI.[45] Normally, the posterior tuberosity of the astragalus abuts against the posterior rim of the tibia and acts as a block to complete plantar flexion. By resecting a sufficient wedge from the anterior end of the astragalus, the tarsus may be displaced dorsally, so that when the astragalus lies in extreme equinus the tarsus

[44] Campbell, W. C : Bone block operation for drop foot, J. Bone & Joint Surg. 12·317, 1930.

[45] Lambrinudi, C.· New operation for drop foot, Brit. J. Surg. 15·193, 1927.

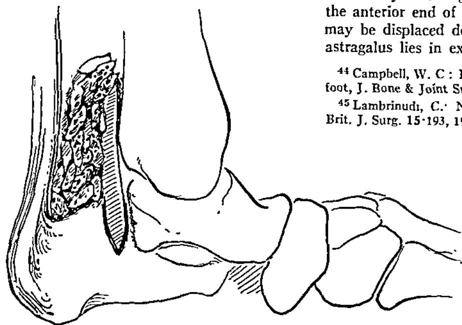


FIG. 159. Campbell's posterior bone block operation for equinus deformity due to paralysis of the anterior leg muscles.

posterior extremity of the os calcis. In addition, the extensor hallucis longus must be transposed to the dorsum of the foot. The tibialis anticus should be transferred by freeing the tendon and the muscle belly up to its middle third (where the neurovascular bundle enters) and passing through an opening in the interosseous membrane.

4. *Loss of Both Invertors and Evertors.* Adequate dorsiflexion cannot be secured by transposition of the extensor hallucis longus alone. Stabilization by the Lambrinudi procedure or a posterior ankle bone block prevents plantar flexion (q.v.).

Whenever the peroneus longus is transferred, elevation of the first metatarsal and hallux equinus are resultant deformities. This can be prevented by suturing the distal tendon stump to the peroneus brevis.

STABILIZATION OF THE FOOT. Obliteration of the subastragalar, the astragaloscaphoid and the calcaneocuboid joints, popularly known as "triple arthrodesis," is necessary to correct instability of the foot due to muscle imbalance. Better stability is afforded if the foot is displaced posteriorly to provide a better lever arm for a weakly functioning calf muscle. Where possible, tendon transposition should be added to re-establish muscle balance. The operation is done only after the age of 10; before that time the structures are mainly cartilaginous. Deformity of the foot, with or without instability, is an indication for a triple arthrodesis. Resection of bone should be wedge-shaped, the base of the wedge being directed toward the convexity of the deformed foot, the apex toward the concavity For example, in equinovarus deformity, the base of the resected bone is superior and lateral, its width corresponding to the degree of deformity. Bony overgrowth generally occurs in the tarsals in the direction of the convexity and must be resected. The final position requires that the heel be in slight valgus. If the deformity be planovalgus, the astragalus plantarflexes, the astragalar head forms a medial and plantar bony prominence, and the longitudinal arch is flattened. After arthrodesis, positioning the os calcis more medially beneath the astragalus supports the latter and restores the arch. Calcaneocavus can be corrected by adequate midtarsal resection, displacing the foot posteriorly, and transferring the tendons of functioning muscles to the Achilles tendon or the os calcis.

PANASTRAGALAR ARTHRODESIS. When leg muscles are too weak to control the foot at the ankle, and foot instability exists, ankle arthrodesis is performed in addition to a triple arthrodesis. The two procedures may be combined at one operation. The astragalus may be completely removed, thoroughly denuded of cartilage and cortex and replaced. However, circulation to the bone is compromised by the latter procedure, aseptic necrosis often supervenes, and many months of abstinence from weight-bearing are necessary before complete replacement occurs. Therefore, ankle fusion and triple arthrodesis should be done as separate operations. Postoperatively, the astragalus should be positioned in equinus, the degree depending upon the height of the heel to be worn. When quadriceps paralysis is present, an added degree of equinus favors hyperextension and stability of the knee.

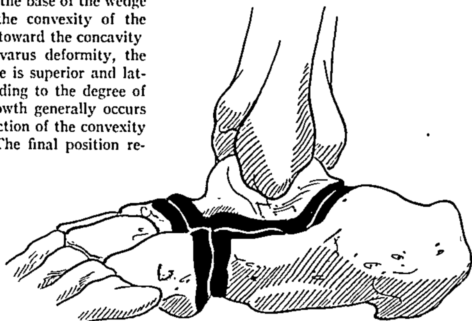


FIG. 158. Triple arthrodesis. The talocalcaneal, the talonavicular and the calcaneocuboid joints are fused.

Fig. 161. Typical poliomyelitic deformity of foot. Weak anterior tibial and tight heel cord. Dorsiflexion ineffectively accomplished by extensor hallucis longus. Cavus deformity developing. Treatment: (1) *correct deformity* by wedge osteotomy of tarsus and plantar fascia stripping; (2) *provide dorsiflexor power* by transferring the extensor hallucis longus to the neck of the first metatarsal and arthrodesing the interphalangeal joint of the big toe.



metatarsophalangeal joint is released, and the dorsal capsule is severed and imbricated. The flexor hallucis longus tendon may be severed at its insertion and fixed to the neck of the metatarsal to act as an active corrective force. Muscle imbalance should be evaluated. An overacting tibialis anticus is attached to the mid-line of the foot. Occasionally, arthrodesis of the metatarsophalangeal joint is desirable.

Cavus Foot and Claw Toes. Clawing of [illegible] teristic polio- [illegible] perextension at the metatarsophalangeal joint and flexion at the interphalangeal joint. It is due to overaction of the toe extensors, the antagonists of the intrinsics. Excessive tension of the extensors is brought about by (1) *weakness of the foot extensors* which allows the forefoot to drop, the toes being drawn up more so by the patient's attempt to dorsiflex the foot by the toe extensors. By passively displacing the forefoot dorsally, the extensor tendons are relaxed, and the toes fall downward to their normal position. (2) *Contracture of the plantar aponeurosis* pulls the forefoot plantarward, the extensor tendons being rendered taut. A high longitudinal arch (cavus) is created. The former mechanism is immediately evident, whereas the latter develops insidiously. If the deformity persists, it is rendered permanent by joint contractures. The depressed metatarsal heads on the plantar surface, the dorsal angulation at the proximal interphalangeal joints, and the tips of the toes pointing downward create points of pressure over which painful calluses form. (3) *Paralysis of the in-*

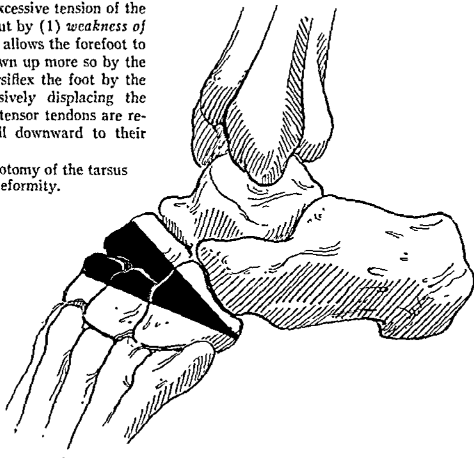


Fig. 162. Wedge osteotomy of the tarsus for cavus deformity.

will lie in a position of a right angle to the tibia. Therefore, a strong triceps surae is a necessary prerequisite.

An oblique incision extends from the front of the lateral malleolus distally to the base of the 3rd metatarsal. The sinus tarsi is cleaned out, and the extensor digitorum brevis is elevated from the os calcis. Capsular structures are excised from the astragaloscaphoid and the calcaneocuboid joints, and cartilage and cortex are removed from the apposing articular surfaces Next, the subastragalar joint is exposed Cartilage and cortex are removed from the superior aspect of the os calcis. Then a large wedge-shaped section is removed from the head, the neck and the body of the astragalus, including cartilage, cortex and cancellous bone. The base of the wedge is directed distally, the osteotome or saw blade emerging at the superior rim of the astragalar head The width of the wedge depends upon the amount of equinus to be overcome. Multiple slivers of cancellous bone are gouged up from all apposing surfaces to afford maximum area contact. Next, the tarsals and the foot are displaced upward so that the scaphoid comes to lie almost completely dorsal to the distal end of the astragalus. A notch cut in the inferior aspect of the scaphoid helps to maintain the displacement. This position is held while the soft tissues are closed and a cast applied.

The Elevated First Metatarsal (Dorsal Bunion). Normally, the anterior tibial muscle, which attaches to the first cuneiform and the base of the first metatarsal, acts to elevate the metatarsal. The antagonist is the peroneus longus. When the latter is weak, the first metatarsal displaces upward. To obtain the push-off in walking, the large toe is flexed actively downward. Therefore, the deformity is most apparent at first only with weight-bearing, but eventually the deformity becomes fixed. The plantar capsule of the metatarsophalangeal joint and the flexor hallucis brevis become contracted. The head of the metatarsal is prominent dorsally where a bony excrescence, the bunion, develops. Removal of the peroneus longus for transfer in the presence of a good anterior tibial is a common cause of the deformity. The anterior tibial should be removed and transferred more laterally about the 3rd cuneiform.

When the dorsiflexors of the great toe are paralyzed and the flexors are intact, the toe is strongly flexed downward to attain stability and push-off. Secondarily the metatarsal displaces upward.

Treatment. Wedges of bone are removed from the metatarsocuneiform and the scaphocuneiform joints with the bases directed plantarward, and fusion is obtained in the corrected position. The plantar capsule of the

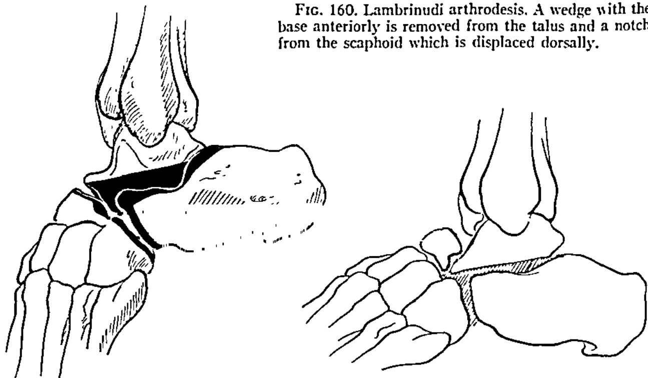
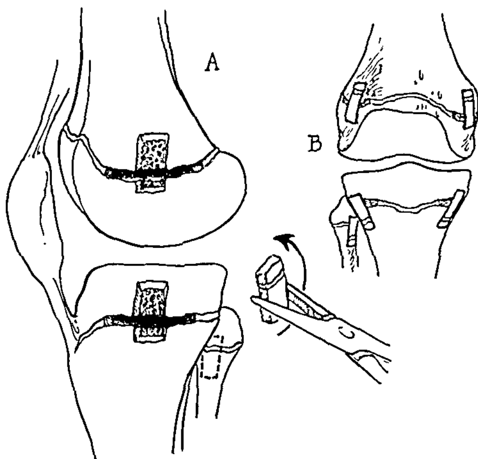


Fig. 160. Lambrinudi arthrodesis. A wedge with the base anteriorly is removed from the talus and a notch from the scaphoid which is displaced dorsally.

Fig. 163. Permanent epiphyseal arrest. The rectangular segment of cortex is removed and reversed so that solid bone bridges the epiphyseal line.



bone.[47] Restriction of growth in the opposite extremity is simpler, safer and more desirable.

Procedure.[48] At the initial visit, accurate leg lengths are determined by roentgenographic measurement. X-ray magnification of 20 per cent must be subtracted to obtain the actual lengths which are measured from the top of the femoral heads to the lower end of the tibiae. The discrepancy is recorded Similar studies are conducted over the next 2 years. The increased discrepancy indicates the rate of lost growth per year. Recognizing that growth halts at 14 in girls and 16 in boys, the yearly decrement in the remaining growth years is added to the already existing shortening to calculate the final anticipated discrepancy. Next, one must determine the appropriate time for epiphyseal arrest. The lower femoral epiphysis contributes ⅜ inch growth per year. The upper and the lower tibial epiphyses each contribute ¼ inch per year. One or more epiphyses may then be closed at a time sufficiently in advance of maturity to make up for the eventual shortening.

Example: At age 8, the female patient has 1 inch of shortening. At age 10, shortening is 1½ inches. Therefore, the decrement is ¼ inch per year. Eventual discrepancy at age 14 will be 2½ inches. Closure of the lower femoral epiphysis will restrict growth ⅜ inch per year over the next 4 years, or 12/8 inch or 1½ inches. Closure of the upper tibial and fibular epiphyses at age 10 will result in a loss of ¼ inch per year over the next 4 years, or 1 inch. Together, total loss following both epiphyseal arrests will equal 2½ inches, thereby balancing the shorter extremity. The actual skleletal age should always be checked by Todd's maturation chart before proceeding. This is a more nearly accurate way of determining the time of maturation.

Permanent Epiphyseal Arrest.[49] The

[47] Abbott, L C., and Saunders, J. B. deC. M : The operative lengthening of the tibia and fibula, preliminary report on further development of principles and technic, Ann Surg. 100 961, 1939.

[48] White, J. W.: Leg Length Discrepancies, Instr. Course Lectures, Am. Acad Orth. Surg , vol 6, Ann Arbor, Mich , Edwards, 1949.

[49] Phemister, D. B.: Operative arrestment of longitudinal growth of bone in the treatment of deformities, J. Bone & Joint Surg 15:1. 1933.

trinsics likewise produces this deformity. As a rule, all 5 toes are contracted. A cavus and claw toe deformity less commonly develops secondary to spina bifida and other diseases of the central nervous system.

Treatment. The deformity can often be prevented by recognizing and treating the causative factor. A drop foot should be supported by a brace. Constant manipulation can overcome the effect of contracture of the plantar aponeurosis. When the deformity is fixed, surgical measures are necessary.

CLAWING OF THE LARGE TOE. The long extensor tendon is severed at its insertion and fixed to the neck of the first metatarsal to supply an active force elevating the head of the bone. The interphalangeal joint is straightened by a wedge-shaped resection and arthrodesed (Jones operation).[46]

CLAWING OF LESSER TOES. The base of each proximal phalanx is removed; the interphalangeal joints are resected, straightened, and fixed with intramedullary pins. This results in some loss of flexor power which does not interfere with walking.

CAVUS (CLAWFOOT). The mild early deformity is little more than an exaggerated longitudinal arch and slight flexion of the toes which is obliterated on weight-bearing. Painful calluses over the plantar aspect of the metatarsal heads disappear when pressure is relieved by a metatarsal pad or a metatarsal bar on the sole of the shoe. Arch strain is overcome by an arch support. Severe deformity requires surgical correction.

Surgical Treatment. When clawed toes are the major deformities, removal of the base of each proximal phalanx allows the toes to drop down. The interphalangeal joints must be straightened and fused. The long extensors may then be transferred to the necks of the metatarsals. An alternative is the removal of the outer 4 metatarsal heads. More commonly, the plantar fascia is thick and contracted, and bony deformity at the tarsus is extreme and fixed. Therefore, it is necessary to resect a bony wedge from the tarsus with its base directed dorsally. At the same time, stripping of the plantar aponeurosis from the plantar surface of the calcaneus is done. Finally, the tarsal area is arthrodesed. Overcorrection must be avoided, as flatfoot or rocker-bottom foot results.

When muscle imbalance is present, and correction is attempted during the growth period, some recurrence of deformity may be anticipated, even though arthrodesis is done. This may be prevented by transferring the toe extensors to the necks of the metatarsals, thereby creating an active dorsiflexing force.

AVOIDANCE OF ACHILLES TENDON LENGTHENING. Dropping of the forefoot, in contrast with the calcaneus which remains in its normal horizontal position, produces an apparent limitation of dorsiflexion at the ankle. By elongating the Achilles tendon, the anterior end of the calcaneus will displace upward and reproduce the cavus deformity. Unless the calf muscles are actually contracted, lengthening of the Achilles tendon is contraindicated.

THE SHORT LOWER EXTREMITY. Poliomyelitis is the commonest cause of retarded growth in the lower extremity. The mechanism is obscure. The reduced rate of growth once initiated continues until bony maturity. Bony maturity in the female generally coincides with the onset of menstruation. A short leg causes pelvic tilt, deviation of the lumbar spine, and a limp. Extra strain is placed on the lumbosacral joints. Equalization of the lower extremities is desirable. However a paralyzed lower extremity requiring a long brace ambulates better with a slight amount of shortening.

Treatment. Conservative methods include the wearing of a high heel or a cork sole elevation in the shoe, tilting the pelvis and walking with a flexed knee on the long side. Surgically, lengthening of the extremity would seem to be ideal. However, the procedure is formidable, requiring extensive freeing of the soft tissues as well as an osteotomy; the major surgery is perpetrated upon an already devitalized extremity; and convalescence is prolonged. Sequelae include (1) foot deformities, (2) knee deformities, (3) bowing of tibial fragments with nonunion or malunion, (4) limitation of motion at the ankle, (5) weakening of leg muscles, (6) nerve complications due to overstretching, (7) circulatory disturbance, (8) infection and (9) aseptic necrosis of

[46] Jones, R.: The soldier's foot and the treatment of common deformities of the foot. II. Claw-Foot. Brit. M. J. 1:749, 1916.

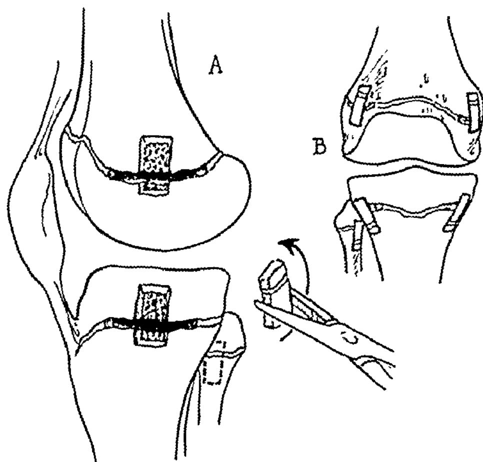


FIG. 163. Permanent epiphyseal arrest. The rectangular segment of cortex is removed and reversed so that solid bone bridges the epiphyseal line.

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Procedure.[48] At the initial visit, accurate leg lengths are determined by roentgenographic measurement X-ray magnification of 20 per cent must be subtracted to obtain the actual lengths which are measured from the top of the femoral heads to the lower end of the tibiae. The discrepancy is recorded. Similar studies are conducted over the next 2 years. The increased discrepancy indicates the rate of lost growth per year. Recognizing that growth halts at 14 in girls and 16 in boys, the yearly decrement in the remaining growth years is added to the already existing shortening to calculate the final anticipated discrepancy Next, one must determine the appropriate time for epiphyseal arrest. The lower femoral epiphysis contributes 3/8 inch growth per year. The upper and the lower tibial epiphyses each contribute 1/4 inch per year. One or more epiphyses may then be closed at a time sufficiently in advance of maturity to make up for the eventual shortening.

Example: At age 8, the female patient has 1 inch of shortening. At age 10, shortening is 1 1/2 inches. Therefore, the decrement is 1/4 inch per year. Eventual discrepancy at age 14 will be 2 1/2 inches. Closure of the lower femoral epiphysis will restrict growth 3/8 inch per year over the next 4 years, or 12/8 inch or 1 1/2 inches. Closure of the upper tibial and fibular epiphyses at age 10 will result in a loss of 1/4 inch per year over the next 4 years, or 1 inch. Together, total loss following both epiphyseal arrests will equal 2 1/2 inches, thereby balancing the shorter extremity. The actual skleletal age should always be checked by Todd's maturation chart before proceeding. This is a more nearly accurate way of determining the time of maturation.

PERMANENT EPIPHYSEAL ARREST.[49] The

[47] Abbott, L C, and Saunders, J B deC. M.: The operative lengthening of the tibia and fibula, preliminary report on further development of principles and technic, Ann Surg 100 961, 1939

[48] White, J W Leg Length Discrepancies, Instr. Course Lectures, Am Acad Orth Surg, vol 6, Ann Arbor, Mich, Edwards, 1949

[49] Phemister, D B: Operative arrestment of longitudinal growth of bone in the treatment of deformities, J. Bone & Joint Surg. 15:1, 1933.

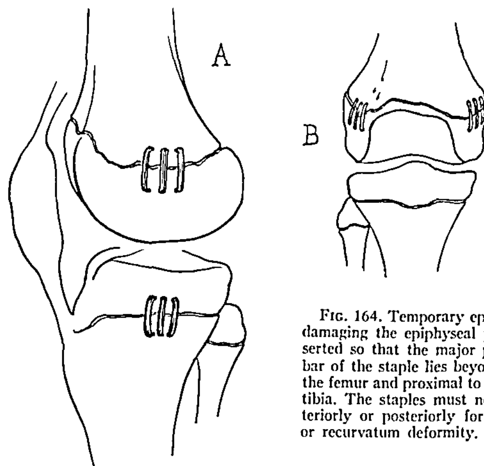


Fig. 164. Temporary epiphyseal arrest. To avoid damaging the epiphyseal plate, the staples are inserted so that the major portion of the transverse bar of the staple lies beyond the epiphyseal line of the femur and proximal to the epiphyseal line of the tibia. The staples must not be placed too far anteriorly or posteriorly for fear of creating flexion or recurvatum deformity.

lower femoral epiphyseal plate is exposed on the lateral side in the interval between the vastus lateralis and the lateral intermuscular septum. The lateral geniculate vessels are ligated, cut and retracted. The cartilaginous plate presents as a white line which passes obliquely from above posteriorly to downward and forward anteriorly. A rectangular block of bone is removed midlaterally, crossing the plate, and includes 1 cm. of epiphysis and 2 cm. of diaphysis. The disk is curetted or chiseled out anteriorly and posteriorly to a depth of several centimeters. Finally, the block of bone is reversed, reinserted and held by firm closure of the overlying soft tissues On the medial side exposure is made by an inci-



Fig. 165. Stapling operation for genu valgum. The medial epiphyseal plates have been retarded by staples until the desired correction was obtained as shown in the illustration at the right. Correction of the right knee is more satisfactory than the left, but clinically the deformity was overcome.



Fig. 166. Demonstrating assistive and resistive exercises, using springs. These exercise springs may be obtained at a sporting goods store.





Fig. 167. (*Left*) Postpoliomyelitic scoliosis brace [illegible] view. Open front permits [illegible]

lateral compression pads.

sion centered over the adductor tubercle and developed between the vastus medialis and the medial intermuscular septum. The medial geniculate vessels are ligated, divided and retracted. The same procedure is followed as on the lateral side. It must be remembered that the plate is saucer-shaped, its peripheral edges lying proximal to the center. This relationship becomes less pronounced with advancing age.

The upper fibular epiphysis is exposed by an incision over the head of the fibula. The common peroneal nerve should be isolated and protected. The cartilaginous disk is completely curetted out, and the defect is filled with small bone chips. Through the same exposure, the upper tibial epiphysis is exposed by elevating the origin of the extensor muscles. Medially, the approach is along the anterior margin of the sartorius tendon. After removal of a rectangular block of bone from both sides of the tibia, the cartilaginous plate, which is in the form of an inverted saucer or V, is curetted out. Each block is reversed and reinserted.

Postoperatively, immobilization for 3 weeks is required. If removal of the plate is inadequate in any one portion, growth may continue therein with consequent deformity.

TEMPORARY EPIPHYSEAL ARREST.[50, 51] Haas found that by encircling the epiphyseal plate with a loop of wire epiphyseal growth is arrested. When the wire is removed, normal growth is resumed. Blount confirmed this work but used staples instead of wire. When sufficient staples are inserted so as to bridge the epiphyseal plate, cessation of growth is immediate and complete. Growth pressure is tremendous, so that one or two staples usually prove to be inadequate and will bend or break. To halt growth completely, 3 staples are inserted on each side of the epiphyseal plate. After sufficient shortening has been attained and limb lengths equalized, the staples are removed, and normal growth is resumed. This method dispenses with the necessity of calculating the appropriate time for epiphyseal closure and the uncertainties of factors of growth and maturation. Premature epiphyseal closure is a rare complication. Stapling can be used to correct deformity. For example, a knock knee can be overcome by temporarily restricting the medial end of the lower femoral epiphyseal plate.

LOWER EXTREMITY INEQUALITY AFTER CESSATION OF GROWTH. After the epiphyses have fused, surgical shortening of the femur or the tibia is indicated. Femoral shortening is preferable because the bone is deeply situated beneath the musculature where deformity due to the bone graft and overlap of fragments is not visible, delayed union or nonunion is



FIG. 169. Postpoliomyelitis. Scoliosis, weak abdominals, weak intercostals, pelvic tilt, iliotibial band contracture.

[50] Blount, W P, and Clarke, G R Control of bone growth by epiphyseal stapling, J Bone & Joint Surg. 32A:464, 1949.

[51] Haas, S L: Retardation of bone growth by a wire loop, J. Bone & Joint Surg 27:25, 1945.

FIG. 170. The Hubbard tank.

FIG. 171. Showing, from left to right, parallel bars, crutch walking, stall bars, floor exercise mat and bicycle resistance exerciser.

Fig. 172. The Elgin table (*left*), demonstrating assistive abdominal exercises. The tilt or stand-up table (*right*) is used to regain balance in standing.



Fig. 173. The Elgin table. By a system of cables and weights, assistive and resistive exercises are performed.

rare, and thigh muscles quickly recover their strength. The femur may be osteotomized obliquely, overlapped and held by screws. A bone graft ensures union. If loss of length is obviously confined to the tibia, shortening of the tibia may be preferable. A step-cut osteotomy of the tibia is performed. The removed segment of bone is used as a bone graft about the osteotomy site. A comparable segment of fibula is also removed.

GUILLAIN-BARRÉ SYNDROME
(Encephalomyeloradiculitis)

Definition. Guillain-Barré syndrome is an acute disease characterized by radicular pain, reversible flaccid paralysis, and a high spinal fluid protein and low cell count.

Etiology. Unknown.

Pathology. Degenerative changes occur throughout the central nervous system but chiefly in the peripheral nerves. Myelin disintegration is especially observed in perivascular areas.

Clinical Picture. The disease starts acutely in an adult, often following a respiratory infection. The patient is afebrile. Often the initial complaints are pain and paresthesia in the extremities, muscle soreness and tenderness, and progressively increasing weakness, beginning in the feet and spreading to involve all 4 extremities. If the disease extends further, the medullary centers become involved, causing bilateral facial paresis, aphonia, dysphagia and dysarthria.

On examination, a flaccid weakness or paralysis and diminished deep reflexes are found. Superficial sensation is reduced over an area encompassing an entire extremity or part of an extremity. The course is generally too short to permit change in electric reaction or development of atrophy.

Laboratory Findings. The total protein in the spinal fluid is elevated as high as 800 mg. Cell count, serology and chemistry are normal. As improvement takes place, the protein content returns to normal.

Treatment. The outlook is excellent for complete recovery. The possibility of bulbar involvement must be recognized, so that early tracheotomy and the use of the respirator are lifesaving. Otherwise, treatment is mainly supportive by tube feeding, transfusions, antibiotics, etc., until the paralysis subsides. Residual weakness is minimal and rare.

CEREBRAL PALSY

Cerebral palsy is a state of muscular dysfunction which results from injury or disease of the upper motor neurons at the level of the cerebral cortex or throughout the course of their fibers within the brain. The loss of inhibitory control results in excessive impulses emanating from lower motor neurons. Lesions of the cerebellum are included, causing symptoms of ataxia and in-co-ordination.

Etiology

The following are the chief factors:

1. Brain Injury at Birth

A. Mechanical causes are poorly applied forceps, excessive uterine contractions, excessive traction on the neck (ruptured vein causing intracranial hemorrhage), sudden pressure change caused by precipitous extrusion of the fetus. Prematurity is a contributing factor, because the delicate vessels beneath the anterior fontanel are easily ruptured, and the leg areas of the motor cortex are damaged by hemorrhage with consequent *spastic hemiplegia.*

B. Prolonged anoxia may be due to excessive use of analgesics and anesthetics, umbilical cord wrapped around the neck of the fetus, tracheal obstruction. The nerve cells are highly susceptible to anoxia.

2. Congenital Brain Defects. Disease in mothers during the first 3 months of pregnancy is associated with a high incidence of congenital anomalies. Diffuse and symmetric involvement by ataxia and athetosis is the usual resultant clinical picture.

3. Rh Factor. The severe jaundice of erythroblastosis fetalis damages the basal ganglia, resulting in athetosis.

4. Postnatal causes include *encephalitis, convulsions* and *head trauma.*

Typical Clinical Picture

A history is often obtained of a difficult birth or of illness during the early months of pregnancy. The infant is late in sitting up, standing, walking and talking. The face is expressionless and may exhibit grimacing and drooling. Speech is difficult. Motion is clumsy, slow, jerky and un-co-ordinated. The shoulder

is adducted and rotated internally, the elbow flexed, the forearm pronated, the wrist flexed and deviated ulnarward, the fingers flexed, and the thumb adducted into the palm. The hip may be flexed, adducted and rotated internally, the knees flexed, the ankle plantar flexed, the foot in equinovarus, and the toes flexed. The child must be supported under the armpits when standing, and the lower extremities are held tightly pressed together or crossed in scissorslike fashion, and the heels cannot be brought down to the floor when standing. The gait is un-co-ordinated. Mass movements occur, i.e., attempts to move one portion of an extremity throws all the other muscles into a state of spasm. Passive attempts to move a joint are resisted by a spastic group of muscles which may respond reflexly by a strong sustained contraction Clonus is often seen. The deep reflexes are hyperactive, and pathologic reflex response is obtained. Athetosis may gradually appear within the first 2 years. Mental retardation becomes evident during the 2nd and the 3rd years. The clinical picture varies with the location and extent of the lesion.

Clinicopathologic Types

The location of the lesion determines the predominating clinical symptoms and findings. Although the following specific types are identified, a mixture of types is usual, but the most outstanding symptoms generally classify the offending lesion.

1. **Cerebral Cortex Lesion**

A. Premotor Area. *Spasticity* results and is evidenced by increased muscle tone, exaggerated deep tendon reflexes, clonus, pathologic reflexes and the stretch reflex. The stretch reflex of a muscle is elicited by stretching that muscle by passively bending or extending the joint over which the muscle acts. The spastic muscle will react by an abnormally strong contraction.

B. Motor Area. *Flaccidity* results and is evidenced by decreased muscle tone, diminished deep tendon reflexes, and abnormal elongation of the muscle, which can be stretched without evoking a stretch reflex. A flaccid muscle must be differentiated from a normal muscle which is weak as a result of stretching and lack of use. A cerebral flaccid muscle may be made to contract by "confusion" on resisted contraction of another muscle. For example, cerebral flaccid dorsiflexors of the foot will contract when active flexion of the hip is resisted.

Hemorrhage varies in extent and location. It may affect both motor and premotor areas so that a mixture of spastic and flaccid muscles result. If the lesion involves the vertex, the legs are affected; if lower on the cortex, the arms are affected; if on the dominant side of the brain, the speech area is affected.

2. **Basal Ganglia Lesion.** Athetosis results, as evidenced by irregular, arrhythmic, involuntary movements, chiefly of the hands, accentuated by voluntary effort and emotion, and subsiding during sleep. The face is expressionless, but involvement of facial muscles causes constant grimacing and twitchings. In his effort to control involuntary movements, the patient increases muscle tension, which should not be confused with spasticity. The stretch reflex is absent. Athetosis when controlled in one region by braces or surgical fixation will reappear in another area of the extremity.

3. **Cerebellar Lesion.** Characteristic signs *of cerebellar dysfunction include ataxia, loss* of sense of balance, muscle in-co-ordination, adiadochokinesia, nystagmus and dizziness. The usual cause is a congenital defect or, less commonly, a hemorrhage at birth. Often the ataxia improves spontaneously as the patient learns voluntary control of balance.

4. **Diffuse Brain Damage (Prolonged Anoxia, Multiple Petechial Hemorrhages, Encephalitis).** Generalized *rigidity* of muscles results and is manifest by loss of muscle elasticity and a "lead pipe" resistance to passive flexion and extension of a joint. The degree of rigidity varies from time to time. No true stretch reflex is present, nor are the deep reflexes hyperactive. Usually the *mentality is deficient*. Neurectomy is valuable for true spasticity but is of no value for rigidity.

Evaluation of Mental Status

About 70 per cent of cerebral palsy patients have a mentality within normal limits. The speech difficulty should not be interpreted as a sign of mental deficiency. Rather, the response to tests made on several occasions should determine the degree of intelligence. Often a child becomes aware of his physical handicap and inability to play with other

children, and development of mental faculties is retarded.

Treatment

Rehabilitation Program. About one third of patients are feeble-minded, and another third are crippled severely and irremediably. These require institutional care. The remainder can be rehabilitated by re-education, which includes training in balance and posture, locomotion, relaxation, rhythmic exercises and speech.

At first only fundamental active motion is taught. This means acquiring the earliest primitive motion of the infant, who at first reaches out with one hand to grasp an object, later with both hands. First, one leg is kicked; later, both legs. These voluntary rudimentary exercises are performed before more complicated motions. The aim should be toward developing the weak antagonists of spastic muscles At the same time the spastic muscles are stretched repeatedly but gently to avoid exciting the stretch reflex. Exercises are performed rhythmically and with increased speed to develop co-ordination. This can be effected by having the child relax on the floor and perform movements to the accompaniment of music. Constant repetition enables the patient eventually to develop these actions without interference by the stretch reflex.

Temple Fay Method.[52] This procedure is aimed at the development and the organization of automatic spinal reflexes as observed by Sherrington and Babinski in the decerebrate animal, and the ultimate co-ordination of these reflexes with what remains of higher cortical control At first, the patient is considered as an amphibian, i.e., one who because of partial or complete loss of cortical control must depend upon the midbrain It is usually simple to teach the fundamental motion excited at this level. The child is placed prone with the chin forward on a polished floor. First, the arm and the leg of one side is made to flex, the extended thumb pointing toward the face, which is turned toward the hand. As the limbs are extended outward and downward, the limbs of the opposite side flex, the head rotating toward that side. A regular rhythm of alternating movement is kept up to the accompaniment of music until a definite pattern is developed. The original jerky movements become replaced by a smooth series of regular muscular contractions. Once this homolateral or amphibian pattern is well developed, the child progresses to the reptilian or next stage of evolution, characterized by the crossed or contralateral pattern of movements. As the right arm flexes and the head rotates toward that side, the opposite leg flexes. Eventually, these crossed movements can be performed without passive assistance. After these motions are established, further advanced patterns are developed through stages of creeping and crawling to independent walking, feeding and writing.

Contractures develop early in the presence of spasticity. Foot equinus is common and can be prevented by repeated stretching of the Achilles tendon and wearing a night brace to maintain dorsiflexion. The brace must be worn throughout the growth period, because the taut calf muscles do not keep pace with the growth in length of the tibia. Also, the hips and the knees must be stretched to prevent contracture. Full-length braces for the lower extremity must be avoided, as they prevent motion and development of weakened and stretched antagonists. Occasionally, this may be used as a temporary measure to enable the patient to gain a sense of balance. The arm is stretched to prevent and overcome adduction contracture. Wrist and fingers are stretched and put through a full range of motion. Contractures do not develop in the athetoid because of constant motion. Because purposeless movements do not appear during sleep, treatment is aimed at relaxation, not repetitive exercises. Braces can be utilized to control athetosis of the lower extremities. Unfortunately, surgical obliteration of a joint affected by athetoid motion will result only in the appearance of involuntary motion elsewhere in the extremity.

The treatment of the ataxic is by training in balance, gait and eye-to-hand skill, but is difficult and often unrewarding. The patient with generalized rigidity and mental impairment requires institutional care.

The most valuable nonsurgical procedures[53]

[52] Pollack, G. A · Treatment of cerebral palsy, J Bone & Joint Surg 34B·154, 1952.

[53] Hipps, H. E.: Evaluation of non-operative methods in cerebral palsy, J. Bone & Joint Surg. 30A.695, 1948.

in cerebral palsy are: (1) *bracing*, which enables the spastic or the athetoid to walk between parallel bars and later with crutches; (2) *co-ordination exercises*, e.g., building blocks, modeling clay, ladder climbing, bicycle riding; (3) *diversion exercises*, by which exercises are performed rhythmically to the accompaniment of music; and (4) *repetitious unaided individual efforts*.

DRUGS.[54] The following are in most common use:

Hyoscine, especially in postencephalitic cases. It favors restoration of previously learned automatic motions. It is of no use to the child handicapped from birth, because he has not learned these automatic movements. For example, hyoscine is administered prior to redeveloping reciprocal motion of the legs in walking.

Prostigmin (or neostigmine),[55] which decreases muscle spasm and allows an increased range of motion. It facilitates correction of contracture by braces or wedging casts and aids in obtaining reciprocation by antagonist muscles by lessening the stretch reflex. Speech difficulty is aided by reducing tongue spasticity.

Hydantoin group of drugs, e.g., Dilantin Sodium, are relaxants useful for tension athetosis.

Curare has a temporary relaxing effect in athetosis. The drug is being used experimentally.

Surgical Treatment. PRINCIPLES. Surgical treatment is considered as an adjunct to conservative treatment Contractures are basically caused by overactive spastic muscles which yield to stretching and by strengthening weakened antagonists. Surgery is indicated for correcting persistent deformity, restoring muscle balance and stabilizing the joints of the foot and the wrist. The program is directed chiefly to restoring ambulation. The type of surgery required is established by determining not only the degree of spasticity of muscles but also the presence of muscle power or flaccidity in their antagonists. Preoperatively, bracing the part upon which surgery is contemplated will determine the benefit that one may reasonably anticipate by operative means Surgery is contraindicated in the athetoid and the mental defective. If athetoid muscles are operated upon, the involuntary motions will shift to other muscles of similar function.

NEURECTOMY.[56] Theoretically, resection of a portion of nerve fibers innervating a spastic muscle will weaken that muscle to a degree proportionate to the extent of denervation. Ideally, the residual muscle power should balance the strength of the antagonist. If the antagonist is flaccid, neurectomy is contraindicated. The antagonist muscle weakened by stretching and disuse should be strengthened by appropriate exercises before determining the extent of neurectomy. Muscle balance must be restored. The original Stöffel operation consisted of exposing the main nerve trunk which sends branches to the spastic muscle; the motor nerve tract which supplies the muscle is isolated by electric stimulation and is resected. A simpler procedure is to expose the nerve branches at their point of emergence from the nerve trunk or at their entrance into the muscle. Before neurectomy, a complete study is made of the muscle and its antagonists before deciding on the extent and the method of correction.

Lower Extremity

FOOT DEFORMITY.[57, 58] The spastic exhibits various deformities, including (1) equinus, (2) varus, (3) valgus, (4) calcaneus and (5) flexion contracture of the toes. For a specific deformity, the status of deforming muscles and their antagonists must be established before treatment can be determined.

SPASTIC EQUINUS of the foot and the ankle is the most frequent deformity. Usually, it is due to a spastic, shortened triceps surae. In the growing child it is corrected by a dynamic brace which gradually obtains complete dorsiflexion by a spring, a control dial at the ankle, or by bending the upright lateral bar at intervals. At first, the brace is worn day and night until the foot can be brought flat upon the floor The heel is elevated to avoid exciting the stretch reflex. Thereafter, the brace is worn

[54] Phelps, W. M.: Recent significant trends in the care of cerebral palsy, J South. M A, Feb, 1946

[55] Kabat, H., and Knapp. J.A.M.A. 122 989, 1943

[56] Stöffel, A The treatment of spastic contracture, Am J Orth Surg 10 611, 1912

[57] Campbell's Operative Orthopedics, ed. 2, vol 2, p 1465, St Louis, Mosby, 1949.

[58] Baker, L. D.: A rational approach to the surgical needs of the cerebral palsy patient, J. Bone & Joint Surg 38A:313, 1956.

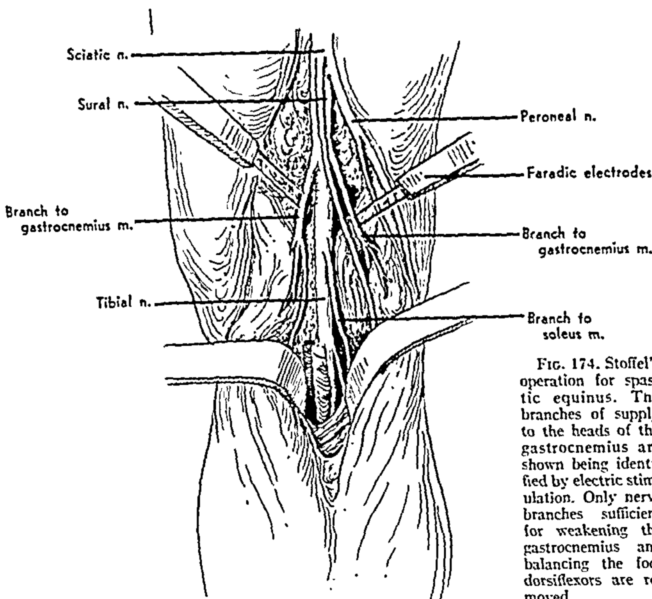


FIG. 174. Stoffel's operation for spastic equinus. The branches of supply to the heads of the gastrocnemius are shown being identified by electric stimulation. Only nerve branches sufficient for weakening the gastrocnemius and balancing the foot dorsiflexors are removed.

only at night for the duration of the growth period. Otherwise, recurrence is inevitable, because growth in length of the tibia bowstrings the calf muscles and the foot naturally assumes the equinus position during sleep. After growth has been completed, deformity requires surgical correction, the procedure depending on the type of muscle imbalance:

1 *Spastic Calf Muscles—Spastic Dorsiflexors.* Overcome deformity by braces. Neurectomy or Achilles tendon lengthening is contradindicated, because a reverse deformity would ensue.

2. *Spastic Calf Muscles—Normal Dorsiflexors.* Treatment consists of partial neurectomy to the gastrocnemius or the soleus, depending upon which is spastic. Spastic involvement of either muscle is determined by exciting the stretch reflex or clonus while the knee is flexed and again extended. The gastrocnemius which attaches above the knee, exhibits a stretch reflex and clonus only while the knee is extended but not when relaxed by knee flexion. The soleus which originates below the knee is unaffected by changes in knee position.

3. *Spastic Calf Muscles—Flaccid Dorsiflexors.* A Lambrinudi type of tarsal stabilization or a posterior ankle bone block overcomes the equinus. Partial neurectomy in addition is necessary only for troublesome clonus or extreme spasticity.

4. *Normal Calf Muscles—Flaccid Dorsiflexors.* The Lambrinudi procedure is indicated. Normal calf muscles are necessary to the success of the operation.

5. *Flaccid Calf Muscles—Flaccid Dorsiflexors.* Panastragalar arthrodesis corrects the equinus and stabilizes the foot and the ankle.

CALF MUSCLE NEURECTOMY. General anesthesia must be light to avoid overcoming the stretch reflex; or local anesthesia may be used. A transverse incision over the popliteal space may be extended proximally at one end and distally at the other to afford the maximum exposure. The tibial nerve lies immediately beneath the deep fascia and superficial to the popliteal vessels. The most proximal branch is cutaneous. Below this, two branches arise, one to the medial and the other to the lateral head of the gastrocnemius. Each divides into several twigs before penetrating the muscle. Immediately below these branches arises a nerve which divides to supply each head of the soleus. The tibial nerve then passes distally beneath the soleus to which it supplies an additional branch. Each branch is tested by faradic current while the foot is held forcefully dorsiflexed. The specific muscle responsible for spasticity and clonus is thus determined, and the branches of supply are avulsed from the muscle or thoroughly excised. Postoperatively, a night brace is worn, and the dorsiflexors are exercised.

ACHILLES TENDON LENGTHENING. The tendon is lengthened only to allow sufficient, but not excessive, dorsiflexion. If it can be definitely established that the gastrocnemius alone is the deforming factor, lengthening of that muscle alone is indicated. The Vulpius procedure consists of transversely dividing the aponeurosis of the gastrocnemius and forcing the edges apart by dorsiflexing the foot.[59]

VARUS AND VALGUS DEFORMITIES are the result of muscle imbalance between invertors and evertors. However, a spastic muscle when transferred will not function sufficiently to restore balance. Nevertheless, before correction of the deformity by bone resection, removal of the deforming influence by neurectomy or tendon lengthening of the tibials or the peronei is essential, and an attempt is made to correct the deformity by progressive wedging with casts. If it can be established that the peronei are normal and the tibials nonfunctioning, tendon transfer may be attempted. When the peroneus longus is transferred, the distal stump should be sutured to the peroneus brevis.

CALCANEUS DEFORMITY occurs as a result of unnecessary or excessive Achilles tendon lengthening or after neurectomy of the calf muscles in the presence of strong or spastic dorsiflexors. Correction of mild deformity may be possible by partially neurectomizing the dorsiflexors and shortening the Achilles tendon. Otherwise, a panastragalar arthrodesis or astragalectomy is necessary.

FLEXION CONTRACTURE OF THE TOES occurs at the metatarsophalangeal joints and is due to spasticity of the intrinsic muscles. The motor branch of the lateral plantar nerve should be resected[60] and plantar capsulotomies done; or the base of the proximal phalanges may be resected. Flexion contracture of the large toe due to overpull of the flexor hallucis brevis is overcome by sectioning of the two heads of insertion of the muscle and cutting the plantar capsule.

KNEE DEFORMITY. *Flexion deformity* is frequent and generally is due to spastic hamstrings in the presence of stretched and weakened quadriceps. The hamstrings must be weakened by a partial neurectomy or tendon lengthening and the quadriceps strengthened by graduated exercises. If the patellar tendon has become elongated from prolonged stretching (indicated by a high riding patella), it must be shortened by transferring the tibial tubercle distally or by plicating the tendon. If flexion deformity is caused by normal hamstrings overpowering a weakened quadriceps, the biceps and the semitendinosus are transferred to the patella. Occasionally, a spastic tensor fascia femoris may be a deforming force through the iliotibial band. Resection of the iliotibial band and, medial to it, the intermuscular septum is necessary. It may also be necessary to cut the heads of origin of the gastrocnemius. This permits further extension of the knee and dorsiflexion at the ankle.

Hyperextension deformity is due to (1) a spastic quadriceps in the presence of normal or weak flexors, (2) equinus deformity of the foot, and (3) excessive hamstring tendon lengthening or ill-advised tenotomies. The first

[59] Vulpius, O., and Stöffel, A.: Orthpadische Operationslehre, Stuttgart, Enke, 1920

[60] Burman, M.S.: Spastic intrinsic muscle imbalance of the foot, J. Bone & Joint Surg. 20:145, 1938

is treated by partial neurectomy; the second, by correcting the equinus; the third, by shortening or reattaching the hamstring tendons. A persistent genu recurvatum is associated with backward bowing of the tibia at the upper end. A wedge osteotomy overcomes the bony deformity.

HAMSTRING TENDON LENGTHENING is performed by exposure of the tendons while the knee is forcefully extended. Each tendon is cut obliquely, permitting further extension of the knee, and the tendon ends are overlapped and sutured together. Occasionally, posterior capsulotomy and cutting both heads of the gastrocnemius also may be necessary.

PARTIAL SCIATIC NEURECTOMY. The nerve is exposed through a longitudinal incision which starts immediately below the gluteal crease midway between the greater trochanter and the ischial tuberosity and extends distally for several inches. The long head of the biceps which originates at the ischial tuberosity and crosses the nerve obliquely is retracted medially. A large branch from the medial side of the sciatic nerve divides into 3 branches, passing to the long head of the biceps, the semitendinosus and the semimembranosus. The selected nerve or nerves are avulsed from the muscles, and the parent branch is resected at its point of emergence from the main nerve. Below this level emerges the branch to the short head of the biceps. This nerve may be left intact to preserve some flexor power.

PATELLAR ADVANCEMENT OPERATION.[61] The patella may be drawn distally and held with a braided wire which passes transversely through the quadriceps aponeurosis immediately above the bone The two strands are threaded distally in the capsule alongside the patella, emerge through the skin beyond the knee, are pulled taut and are tied over a dressing. A pull-out wire is attached proximally. The patellar tendon is plicated and sutured. Postoperatively, the knee is splinted in extension for several weeks, after which the wire is extracted, and gentle motion is begun.

Removal of the tibial tubercle for displacement distally should be avoided, as it may endanger the anterior epiphyseal plate with consequent recurvatum deformity.

[61] Chandler, F A · Patellar advancement operation; a revised technic, J Internat. Coll Surgeons 3:433, 1940.

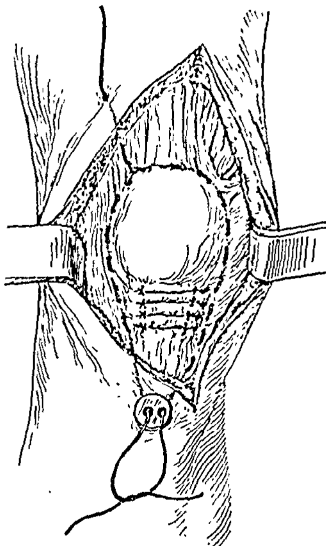


FIG. 175. The patellar advancement operation. The tension on the plicated patellar tendon is relieved by an encircling suture of braided stainless steel wire which holds the patella distally. When the patellar tendon has consolidated, the tension suture is removed by a pull-out wire.

HIP DEFORMITY. In the order of frequency, the deformities are:

Flexion and internal rotation caused by spasticity of the internal rotators, chiefly the tensor fascia femoris and the anterior fibers of the gluteus medius and minimus. It produces the characteristic "scissors" gait wherein the legs cross each other when standing and walking. At first the deformity is only apparent when these muscles are active and can be reduced passively. Later the muscles and the fascia become contracted, and the deformity becomes fixed.

To correct spastic internal rotation, the origin of the tensor fascia lata is transferred from the anterior to the lateral portion of the iliac crest to convert it into an abductor and an external rotator. The gluteus medius and minimus should not be disturbed, as this would seriously weaken abduction In addition, the iliotibial band may require resection. Release of the sartorius and the rectus femoris at their origins corrects flexion deformity. Postoperatively, the extremity is immobilized in external rotation and extension. If internal rotation is fixed, it should be corrected by supracondylar osteotomy. The upper fragment is held in internal rotation by a pin inserted transversely into the bone, the pin being incorporated in the cast, while the distal fragment is externally rotated and properly aligned before inclusion in the cast.

Adduction caused by spasticity of the adductors is uncommon. The legs are held firmly together but *do not cross* as in the scissors gait. It is recognized by the fact that the extremity does not internally rotate when the legs are forced apart. If the gracilis is the major deforming factor, flexion deformity of the knee is associated because the muscle spans the knee on its way to its insertion To test for gracilis spasticity and shortening, the knee is extended passively, whereupon the limb adducts spontaneously.

Treatment consists of paralyzing the adductors longus and brevis by sectioning the anterior branch of the obturator nerve If this proves to be inadequate, the posterior branch may also be removed (For surgical anatomy, see description of the obturator nerve.) The obturator nerve may also be secured by the intrapelvic approach.

Technic Through a Pfannenstiehl incision, the sheath of the rectus is split longitudinally, and the muscle is retracted medially. The properitoneal fat is exposed. By blunt dissection the posterior surface of the rectus is followed to its attachment to the pubis, then along the lateral wall of the pelvis, the bladder and the peritoneum being displaced upward. The nerve is located as it penetrates the obturator fascia, and a large segment of it is resected After obturator neurectomy, some adductor power is preserved, because the pectineus (femoral nerve) and some of the adductor magnus (sciatic nerve) remain active.

If the gracilis alone is involved, it may be sectioned at the knee and allowed to displace upward.

Dangers of Adductor Neurectomy and Tenotomy. The scissors gait and adduction deformity may be simulated in a patient with paralyzed abductors. In order to attain stability in standing and walking, the extremities are held firmly pressed together. By paralyzing the adductors, the compensation mechanism is lost, and the patient is severely disabled Before weakening the adductors, testing for the strength of abduction is mandatory. The test is carried out after injection of local anesthetic into the adductors whose spasticity might interfere with active abduction.

Flexion as an independent deformity is rare and is usually mild. It is caused by spasticity and contracture of the sartorius and the rectus femoris The iliopsoas is rarely involved by spasticity.

Dislocation occurs in severely spastic cases. Severe pain is associated. If the dislocation is of long standing, the acetabulum fails to develop

Before attempting surgery, obturator neurectomy overcomes adductor spasm, the main obstacle to reduction. Also, the origins of the tensor fascia lata, the gluteus medius and the gluteus minimus may be displaced downward by transferring the iliac crest. Traction is applied until the femoral head lies at the level of the acetabulum; then the hip is arthrodesed. If dislocation is bilateral, a subtrochanteric osteotomy is indicated instead.

Combined Correction of Hip and Knee Flexion.[62, 63] When knee flexion deformity exists, hip flexion is a compensatory attitude assumed to attain balance. By removing the tendons of insertion of the hamstrings and transferring them to the femoral condyles, the powerful force which normally flexes the knee is reduced and instead is utilized to extend the

[62] Eggers, G W N. Division of patellar retinaculum, J. Bone & Joint Surg 32A 80, 1950

[63] ——— *Transplantation of hamstring tendons to* femoral condyles to improve hip extension and decrease knee flexion in cerebral spastic paralysis, J Bone & Joint Surg. 34A 827, 1952

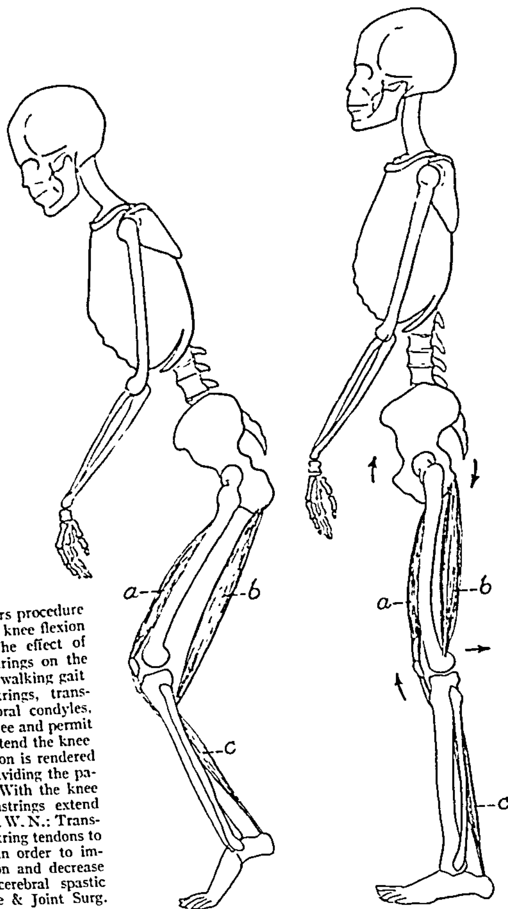


FIG 176. The Eggers procedure for combined hip and knee flexion deformity: (*Left*) The effect of the overacting hamstrings on the tibia in the dynamic walking gait (*Right*) The hamstrings, transplanted to the femoral condyles, no longer flex the knee and permit the quadriceps to extend the knee The quadriceps action is rendered more effective by dividing the patellar retinaculum. With the knee extended. the hamstrings extend the hip. (Eggers, G. W. N.: Transplantation of hamstring tendons to femoral condyles in order to improve hip extension and decrease knee flexion in cerebral spastic paralysis, J. Bone & Joint Surg. 34A:827)

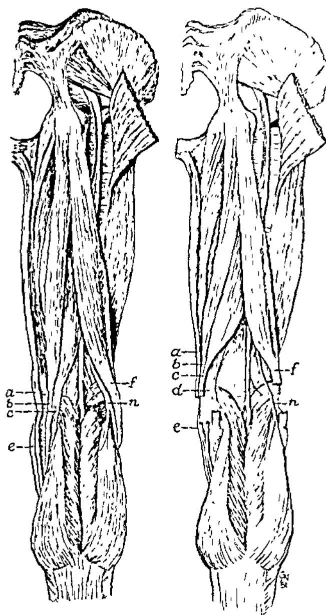


FIG. 177. The Eggers procedure for combined hip and knee flexion deformity. (*Left*) Anatomy involved in transplantation of hamstrings a, gracilis; b, semitendinosus; c, semimembranosus; e, sartorius, f, biceps femoris; n, common peroneal nerve. (*Right*) Showing division of tendons Not divided are d, adductor magnus, and e, sartorius. (Eggers, G W. N.. Transplantation of hamstring tendons to femoral condyles in order to improve hip extension and decrease knee flexion in cerebral spastic paralysis, J. Bone & Joint Surg 34A: 827)

hip joint. By depleting the antagonists, the quadriceps can function at a better advantage. The popliteus, the gastrocnemius and the sartorius prevent knee recurvatum after hamstring transplantation.

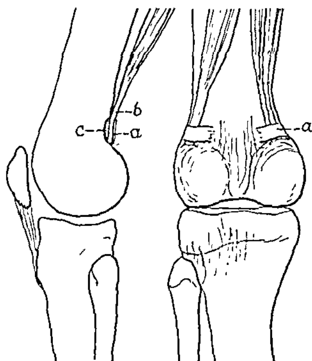
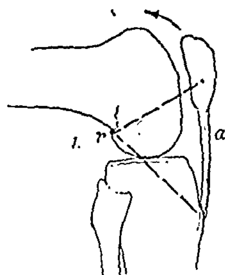


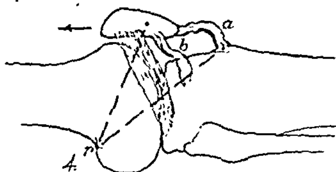
FIG. 178. The Eggers procedure for combined hip and knee flexion deformity, showing subperiosteal placing of tendons. The tendons are buried in an osseous groove: a, periosteal flap; b, tendon; c, osseous groove. (Eggers, G. W. N.: Surgical division of the patellar retinaculum to improve extension of the knee joint in cerebral spastic paralysis, J. Bone & Joint Surg. 32A:80)

Eggers combines hamstring transplantation with division of the patellar retinaculum. The patellar retinaculum may be described as the combined fibers of the fascia lata and the tendinous slips of insertion of the vasti which, in addition to inserting on the patella, send slips to the condyles of the tibia. Prolonged knee flexion causes stretching of the patellar tendon but causes less stretching of the retinaculum medial and lateral to the patella As a result, the quadriceps extensor force is expended upon the retinaculum before pulling the patella upward. By sectioning these lateral fibrous and muscular retinacula, the extensor force is exerted entirely upon the patellar tendon. If the patellar tendon is unusually elongated, it may be necessary, in addition, to advance the patella distally by plicating the tendon or by transferring the tibial tubercle.

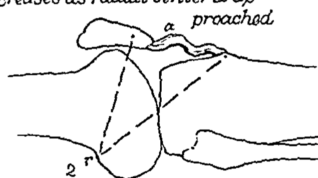
Talipes equinus is seldom present in this type of case. However, if this deformity is present, it may be corrected by a soleus neurectomy.



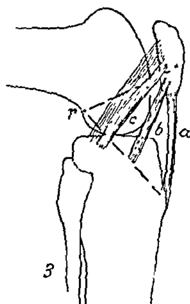
4 Knee extended, quadriceps force is limited by less-stretched muscular & fascial retinacula which are not placed for efficient function and therefore extension is incomplete of the

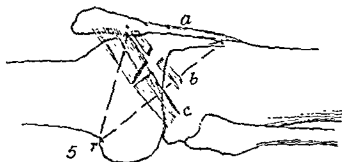


1 Knee flexed, the stretching is greatest at patellar ligament decreases as radial center is approached



2. Knee extended, structures near radial center are taut. The patellar ligament and adjacent structures are not taut.





5. Fascial and muscular retinacula divided, remove restriction and allow quadriceps extensor force to be applied to tibia by patellar ligament.

3. Structures near radial center which are less-stretched are fascial retinaculum and lateral and medial portions of muscular retinacula

a.-patellar ligament
b -muscular retinaculum
c -fascial
r -radial center

Fig. 179. The Eggers procedure for combined hip and knee flexion deformity: diagrammatic representation of patellar extension and the restrictive structures (patellar retinaculum). (Eggers, G. W. N.: Surgical division of the patellar retinaculum to improve extension of the knee joint in cerebral spastic paralysis, J. Bone & Joint Surg. 32A:80)

Technic. Incisions are made medially and laterally over the hamstring tendons. The common peroneal nerve is protected as the biceps tendon is severed at its attachment to the fibular head. Then the tendon is fixed under tension to a groove gouged out from the posterolateral aspect of the lateral femoral condyle. The tendons of the semitendinosus,

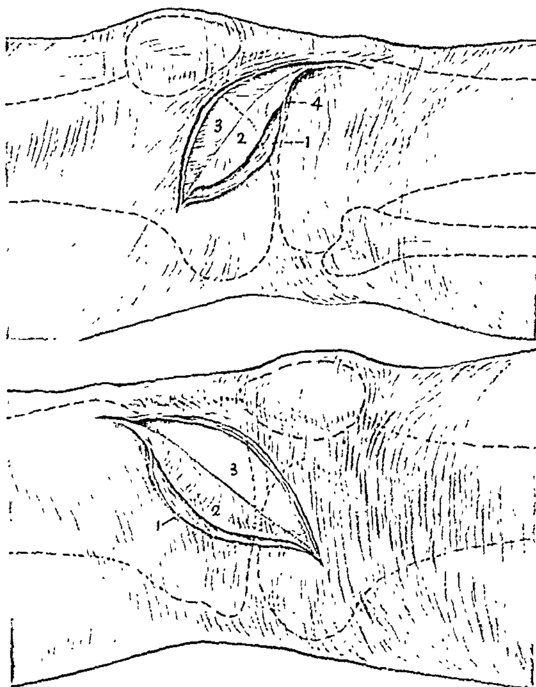


FIG. 180 The Eggers procedure for combined hip and knee flexion deformity: (*Top*) Division of patellar retinaculum, exposure on lateral aspect of knee. Incision must reach level of anterior margin of fibular collateral ligament (1) Fascial retinaculum (2) Muscular retinaculum (3) Capsule. (4) Fusion margin of musculofascial retinaculum. (*Bottom*) Division of patellar retinaculum, exposure on medial aspect of knee. Incision must reach level of anterior margin of tibial collateral ligament. (1) Fascial retinaculum. (2) Muscular retinaculum. (3) Capsule (Eggers, G W. N.: Surgical division of the patellar retinaculum to improve extension of the knee joint in cerebral spastic paralysis, J Bone & Joint Surg. 32A:80)

the semimembranosus and the gracilis are severed at their insertion on the anteromedial aspect of the upper tibia. These are fixed to a groove cut from the posteromedial aspect of the medial femoral condyle.

Next, the patient is placed in the supine position. An incision, 3 inches long is made ½ inch lateral to the lateral patellar margin and parallel with it, beginning at a point midway between the distal end of the patella and the tibial tuberosity. The fascial retinaculum is exposed and divided parallel with the patella and the patellar tendon, and then the incision is curved posteriorly to a point anterior to the femoral attachment of the fibular collateral ligament. The iliotibial band is cut. At a deeper level, the muscular retinaculum is divided in the same line. On the medial aspect of the knee, the incision is similar and is carried backward to the level of the femoral attachment of the tibial collateral ligament. The fascial and next the muscular retinaculum are divided. Postoperatively, the patella lies at a higher level than before. The knee and the hip are immobilized in extension for 3 weeks, and then walking instructions are given.

Upper Extremity. The complex function of the upper extremity makes rehabilitation difficult, because spasticity and in-co-ordinated movement are distributed diffusely. A spastic muscle when transferred functions badly. Therefore, surgery is limited chiefly to tenotomies and, infrequently, neurectomies. Occasionally, flaccidity of a muscle or a muscle group may be interspersed Before a muscle is weakened by tenotomy or neurectomy, the status of the antagonists must first be determined to avoid producing a joint lacking completely in muscular control. When a spastic muscle is balanced by a spastic antagonist, surgery should be avoided, and treatment is confined to an exercise program designed to obtain rhythm and increase the speed of movement.

The most characteristic positions of deformity in the upper extremity are:

1. Finger flexion, wrist flexion and ulnar deviation, forearm pronation, elbow flexion, and shoulder internal rotation and adduction.

2. Hyperextension at the metacarpophalangeal joints, forearm supination, elbow extension, and shoulder external rotation and adduction.

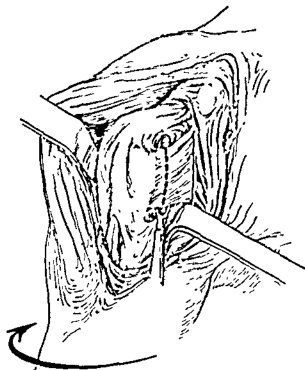


Fig. 181. Sever's operation for adduction and internal rotation deformity of the arm. The tendon of the subscapularis and the upper portion of the tendon of the pectoralis major are severed, and the arm is immobilized in abduction and external rotation.

In dealing with the upper extremity, the shoulder must always be considered first.

Shoulder. Tightness and spasticity of the subscapularis cause internal rotation and adduction. This condition is treated by tenotomy of the subscapularis tendon (Sever's operation). Before this is done, the integrity of external rotation is tested by passively rotating the arm externally and having the patient hold this position against an attempt to internal rotation. If spasticity can be demonstrated in the pectoralis major and the latissimus dorsi, their tendons should be lengthened rather than severed. It is important to retain their stabilizing effect upon the humerus necessary for abduction If the external rotators are spastic, a rotation osteotomy of the humerus reduces the deformity.

The Sever Operation. A longitudinal incision is made over the anterior aspect of the shoulder, starting at the acromioclavicular articulation, extending distally for about 6 inches The deltopectoral cleft is developed, and the cephalic vein is retracted laterally with

the deltoid The tendinous insertion of the pectoralis major is severed, and the muscle is retracted medially. The tip of the coracoid is osteotomized and displaced downward with the attached tendons of the pectoralis minor, the short head of the biceps, and the coracobrachialis. Next, a director is inserted beneath the tendon of the subscapularis, which is sectioned. Good abduction and external rotation are now obtained If contracture is old, section of the tendons of the teres major and the latissimus dorsi is necessary in addition. These tendons are approached best through an incision at the posterior axillary fold. Postoperatively, the extremity is immobilized in an abduction spica cast for 2 to 3 weeks, followed by a brace and active abduction exercises.

ELBOW. When both flexors and extensors are spastic, surgery is contraindicated. When the biceps or the brachialis are spastic and the triceps normal or weak, tendon lengthening of the spastic muscle is done. If the triceps is spastic and the flexors weak, partial neurectomy of the triceps is indicated. Tendon lengthening of this muscle is difficult. Otherwise, an elbow brace with a lock is applied to maintain flexion or extension as desired.

FOREARM. Persistent pronation is corrected if there is sufficient power in the supinators. Tenotomy of the pronator teres is usually sufficient; or the muscle may be divided into several longitudinal strips which are tenotomized alternately at each end and the muscle lengthened by forcefully supinating the forearm.

WRIST. Ulnar deviation is corrected by lengthening the flexor carpi ulnaris where it attaches to the pisiform. To overcome spastic flexion deformity, the flexor carpi ulnaris and radialis or the superficial flexors can be transferred to the extensors. One must be wary of weakening wrist flexion, because the patient may utilize this motion to extend the fingers automatically. Arthrodesis of the wrist in dorsiflexion is permissible only when finger flexors are weak or flaccid and normal wrist muscles can be transferred to reinforce finger flexion. Before obliterating the joint, first a brace or a cast must be applied to establish the effectiveness of the procedure.

HAND. Spasticity of the intrinsics causes flexion at the metacarpophalangeal joints and extension at the interphalangeal joints. The lumbricals can be weakened by stripping them from their origins at the profundus tendons in the palm; or their tendons of insertion to the bases of the proximal phalanges may be severed without disturbing the lateral bands. Spastic extensors and weakened or flaccid lumbricals produce the opposite deformity, namely, extension at the metacarpophalangeal joints and flexion at the interphalangeal joints. The extensors may be weakened by partial neurectomy of the radial nerve branches and the metacarpophalangeal joints flexed by dynamic splinting. Multiple capsulotomies of these joints are usually necessary.

The thumb is usually held flexed, opposed and adducted into the palm, the fingers flexing over it. To correct this deformity, the adductor pollicis is tenotomized, the interphalangeal and the metacarpophalangeal joints are fused, and the thumb is immobilized in volar abduction and moderate opposition. An alternate procedure consists of holding this position by a bone graft wedged between the first two metacarpals (the Thompson procedure). This permits useful pinch and grasp.

Technic.[64] Through a dorsal incision, the cleft is exposed, and the radial artery is identified and protected. A flat bone graft beveled at each end is inserted into a slot made in each metacarpal. These slots are cut so as to lie opposite each other when the thumb is placed in volar abduction and opposition. The adductor tendon may require sectioning. Postoperatively, a cast is applied.

[64] Foerster, O Value of orthopedic fixation in nerve disease, Acta chir. scandinav. 67:351, 1930.

13

Tumors of Bone

Tumors of bone may be divided into (1) those derived from osseous tissues, and (2) tumors composed of nonosseous tissues.[1, 2]

CLASSIFICATION

1. Tumors of Osseous Origin
   - A. Cartilaginous
     - a. Osteochondroma
     - b. Chondroma
     - c. Chondroblastoma
     - d. Chondrosarcoma
   - B. Osseous
     - a. Osteoma
     - b. Osteoid osteoma
     - c. Osteogenic sarcoma
     - d. Parosteal ossifying fibroma
   - C. Resorptive
     - a Bone cyst
     - b. Diffuse osteitis fibrosa cystica
     - c. Fibrous dysplasia
     - d. Giant cell tumor
2. Tumors of Nonosseous Origin
   - A. The Marrow or Haversian Systems
     - a. Ewing's endothelial myeloma
     - b. Multiple myeloma
     - c. Chloroma or leukemia of bones
     - d. Reticuloendotheliosis
     - e. Xanthoma and granulomas of bone
   - B. Metastatic
     - a Carcinoma of thyroid, breast, prostate, kidneys, etc.
     - b. Lymphomas, neuroblastoma, sarcoma
   - C. By Inclusion or Direct Invasion
     - a. Chordoma
     - b Angioma, angiosarcoma
     - c. Fibroma and fibrosarcoma of fascia or nerve sheath
     - d. Myosarcoma
     - e. Synovioma

[1] Geshickter, C. F., and Copeland, M M Tumors of Bone, Philadelphia, Lippincott, 1949

[2] Ewing, J · A review and classification of bone sarcomas, Arch. Surg 4:485, 1922.

HISTOGENESIS

In this author's opinion, the most logical concept of the development of bone tumors is that presented by Geshickter and Copeland. Utilizing this interpretation in the following chapters will clarify the subjects and explain satisfactorily the origin of bone neoplasms This does not imply rejection of other theories. Briefly, the theory may be summarized as follows:

The primitive embryonic mesenchyme differentiates into connective tissue which is capable of developing into fibrous tissue, cartilage, or bone. This mesenchyme condenses at the site of the future skeleton and develops into cartilage at the site of the endochrondal skeleton or into membrane preliminary to bone formation as in the face and the skull.

In membrane bones, spindle cells form bone directly. The process corresponds to osteomas or osteoid osteoma in which bone forms by direct ossification in fibrous tissue. Logically, cartilage is absent.

In endochondral skeleton areas, small rounded fetal-cartilage cells are first formed, intervening matrix is given off, this calcifies, vascular tissue invades, and osteoclasts erode the calcified cartilage, and new bone is laid down The osteogenetic tissue arises from the periosteum and the endosteum. The process is reversible. Throughout life the vascular tissue and the osteoclasts resorb bone in order to supply minerals to the body or preliminary to laying down new bone. This process may become abnormal and a source of tumor formation, as, for example, bone cyst or giant cell tumor. Small islands of cartilage may persist throughout life and later give rise to central chondromas.

The embryonic precartilaginous connective tissue does not disappear altogether. It persists in small islands in synovium, beneath the periosteum, at points of reflexion of joint

capsule, and at points of attachment of tendons and ligaments. This precartilaginous connective tissue is the most common source of bone tumors. This explains the origin of chondromatosis in a joint or the peripheral osteophytes of arthritis. Beyond the joint, where the tendon attaches to a bony prominence, the precartilaginous connective tissue probably is responsible for building these prominences and further length of tendon during longitudinal growth. The tissue is constantly differentiating into fetal cartilage, which is converted into adult cartilage; calcification occurs, then resorption, and finally ossification Unusual activity of this normal process is seen in benign exostoses (osteochondromas). When differentiation is retarded, the process extends only as far as cartilage formation, and very little bone is formed. Cartilaginous tumors result This is especially prone to occur during the growth period If the process is disorderly, the malignant chondromyxosarcoma results. The tissue is unusually active in joint formation about the small bones of the hands and the feet, the vertebrae and the ribs, all of which are affected by central chondromata. After the period of rapid growth, the fetal tissue is a little more advanced, therefore, the process, once initiated, is more likely to produce bone, and more osseous tissue and less cartilage are formed, and the sclerosing form of osteogenic sarcoma becomes manifest by disorderly proliferation and differentiation. By relating bone tumors to this hypothesis, the following concepts are predicated:

1. Cartilaginous growth is represented by chondromas, chondromyxomas and chondromyxosarcomas.

2. Vascularization of calcified cartilage and preformed bone by vessels and giant cell osteoclasts is represented by giant cell tumors and osteolytic osteogenic sarcomas.

3. Ossification in fibrous tissue is represented by osteomas, ossifying fibromas, osteoid osteoma, fibrous dysplasia, myositis ossificans and sclerosing osteogenic sarcomas.

4. Combinations:

Osteochondromas—cartilaginous growth, ossifying fibrous tissue

Chondroblastomas—cartilaginous growth, vascular resorption and giant cell osteoclasts

Bone cysts—vascular resorption, giant cell osteoclasts, healing reaction with ossifying fibrous tissue

OSTEOMA

An osteoma is a benign slowly growing tumor occurring in the membranous bones of the skull and the face.

Clinical Picture

Onset. In childhood.

Predominant Location. Frontal and facial bones. Usually external, but may be intracranial, intranasal, intraorbital, and within a sinus.

Findings. A hard, immovable, moundlike or sessile, nontender swelling over which the soft tissues are freely movable.

Symptoms. External tumors are asymptomatic. Others produce symptoms referable to the part. For example, intracranial osteomas cause epileptic seizures, headache, etc.

Course. Very slow and chronic, tumor reaches a final stationary size. Occasionally, growth may be rapid.

Pathology

The commoner type of *slow growing* osteoma is composed of a dense compact bone covered by a fibrous tissue capsule continuous with the periosteum. The bony structure is continuous with the inner or the outer tables of the skull. The *rapidly growing* tumor is composed of a proliferating vascular fibrous stroma containing newly formed osteoid or osseous spicules. A cancellous structure results. Conspicuous numbers of multinuclear cells are often present and engaged in osseous resorption. This latter type has been called *osteogenic or ossifying fibroma.*[3]

Prognosis and Treatment

Once the osteoma has reached a certain size, it remains stationary. It does not undergo malignant change. Excision is indicated only for symptomatic reasons, rarely for cosmetic. The defect in the skull is replaced by a tantalum plate. The more rapidly growing ossifying fibroma is prone to develop within the spine where pressure symptoms may indicate its removal.

[3] Lichtenstein, L.: Bone Tumors, St. Louis, Mosby, 1952.

PLATE 19. Osteochondroma (× 6). Note the cartilaginous cap and enveloping fibrous capsule.



OSTEOCHONDROMA

This represents the largest group of benign bone tumors and are composed of spongy bone covered by a cartilaginous cap.[4, 5]

Clinical Picture

Age. Osteochondromas appear and grow only during the growth period, rarely thereafter.

Area of Predilection. Metaphyses of long tubular bones, especially about the knee. The remainder develop mainly about the ankle, the hip, the shoulder and the elbow.

Location favors sites of tendinous attachments of strong muscles such as the adductor magnus but may be situated independent of tendons.

Asymptomatic Period of Growth

Symptoms are mild, usually due to an overlying bursitis.

Findings. Firm, nontender swelling which is firmly fixed to bone. Overlying tissue freely movable. The size is palpably larger than is apparent in roentgenograms, because the cap of cartilage is not visualized. The cartilage is often abundant, especially in children. The overlying bursa may be swollen, fluctuant and tender. Limitation of joint motion occurs when the tumor is large and impinges on adjacent structures.

Roentgenologic Findings

The tumor is revealed as an outpouching



FIG 182. Osteochondroma of the hip. The actual size of the tumor is much larger than is apparent on the roentgenogram, because of large, multiple, cartilaginous excrescences which surmount the bony prominence.

[4] Jaffe, H. L: Hereditary multiple exostoses, Arch Path 36 335, 1943.

[5] Schramm, G.: Pathogenesis of cartilaginous exostoses and enchondromas, Arch orthop 27 421, 1929.

Fig. 183. Osteochondroma.



of trabeculated bone at the metaphysis. Its attachment is sessile or pedunculated and composed of cortical and medullary portions which are continuous with cortex and medulla of the tumor and main bone. Externally, the cartilaginous covering is invisible in roentgenograms, unless it calcifies. When sharp definition of the mass is lost and the osseous structure is being resorbed, a stippled or granular appearance results which should arouse suspicion of malignant change.



Fig. 184. Osteocartilaginous exostosis.

Pathology

Gross. A firm, lobulated mass, varying in size, is encapsulated by intimately attached fibrous tissue, which is continuous with the periosteum of the adjacent bone. Removal of the fibrous capsule reveals the shiny smooth cartilaginous covering. In children, during active growth of the tumor, the cartilage is often abundant and lobulated. When growth is halted, the cartilage recedes to a smooth thin layer, which is subject to degeneration and disappearance with advancing age. Beneath the cartilaginous covering, the interior of the tumor consists of cancellous bone and fatty marrow.

Microscopic. The outer covering consists of fibrous connective tissue. Its deepest layer, during active tumor growth, consists of embryoniclike cells, which form sheets of cells differentiating into cartilage cells. As one proceeds toward the deeper levels, one observes formation of matrix between chondrocytes, calcification of the matrix, and invasion by vascular osteogenetic tissue from the marrow. The process of endochondral ossification enlarges the tumor and continues during the growth period of the individual. After growth is halted, the cartilage no longer actively proliferates, endochondral ossification is no longer observed, and the cartilage covering lies on a thin subchondral bone plate. Osteoblasts are rare, and the marrow becomes fatty. The

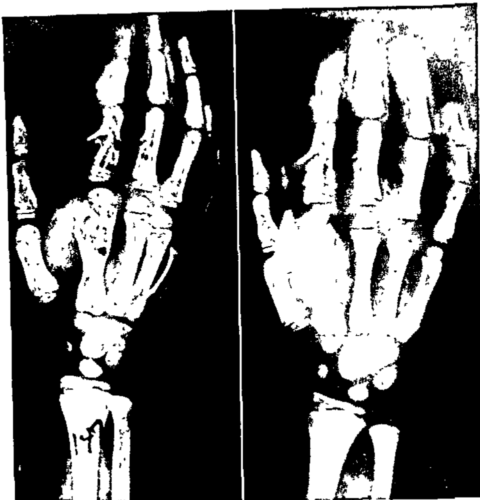


FIG. 185. Enchondroma.

spongy interior is continuous with the spongy bone of the parent bone.

THEORY OF HISTOGENESIS

The deep layer of the periosteum throughout life retains the potential for forming cartilage or bone. The cambium layer produces an embryonic tissue which is the common forerunner for bone and cartilage. The tumor may represent a perverted activity of the periosteum which reverts to its role as perichondrium.

Geschickter and Copeland believe that focal accumulations of embryonic connective tissue are situated at the points of tendinous attachment, thereby explaining the tendency toward tumor formation at these sites.

TREATMENT

Surgical removal is indicated for: (1) interference with joint function, (2) repeated painful bursitis, (3) fracture of the tumor with symptoms and (4) suspicion of malignant change (occurs in 5%). To prevent recurrence, the tumor must be removed at its base. Geschickter and Copeland advise resection of tendon well above its point of attachment in an effort at removal of the precartilaginous connective tissue, thereby preventing recurrence. The tumor is not sensitive to irradiation.

CHONDROMA

(Endochondroma, Chondromyxoma)

A chondroma is a benign tumor of young adults occurring in a central location of bone, usually the phalanx and the humerus, and causing destruction of the cancellous bone. Its main importance lies in its potential for undergoing malignant transformation, particularly when situated in the large long bones or the pelvis.[6]

CLINICAL PICTURE

Age. From 10 to 50

Location. Phalanges of hands and feet. Less

[6] Geschickter, C. F, and Copeland, M. M.: Tumors of Bone, Philadelphia, Lippincott, 1949

commonly the innominate bone and the large long bones

Single lesions, rarely multiple

Symptoms. None or slight soreness. Severe pain occurs acutely from a pathologic fracture or slowly from malignant transformation.

Findings. The phalanx is enlarged as a result of distention of a thinned cortex. In a long bone no deformity is observed, the tumor remaining intramedullary.

Course. Develops very slowly during childhood, then remains stationary unless malignant changes supervene.

Roentgenologic Findings

Roentgenograms show a small translucent loculated or nonloculated area well demarcated from surrounding bone. In a phalanx the cortex is thinned and expanded. In a large long bone the cortex is not involved. The center may exhibit stippling of calcification and striations of fibrous septae. No reactive new bone formation is present. In the small bones of the hands and the feet, the cortex may be perforated, and the shadow of the tumor extends into the soft tissues, although the tumor is benign. On the other hand, erosion of the cortex in a large tubular bone and spread externally strongly suggests malignant change. This is particularly true when the borders of the lesion become mottled and hazy.

Pathology

Gross. The tumor is surrounded by a fibrous capsule which on cut section displays extensions into the interior, dividing the growth into lobules. The neoplastic tissue is composed of bluish-white translucent cartilage, which may contain white areas of calcification, and cysts containing a gelatinous or myxomatous substance. The tumor is not excessively vascular.

Microscopic. The tumor shows stages of formation of cartilage from embryonic tissue The mesenchymal type of tissue is seen only at the periphery of the tumor. As one proceeds toward the deeper areas of the tumor, one observes all stages in the formation of cartilage. The mesenchymal cells become rounded, and matrix accumulates between the cells. The most mature cartilage is found at the center of the tumor where it may undergo the usual degenerative changes of cartilage, namely, calcification, cystic disintegration and myxomatous change. These chondrocytes are normal in appearance. They are small, occur in pairs and tetrads, contain acidophilic cytoplasm, and the nuclei are often small and pyknotic.

In contrast, a chondrosarcoma contains precartilaginous fetal connective tissue everywhere, the tumor is hypercellular, and the cells are irregular in size and contain large, often multiple, dense nuclei.[7]

Diagnosis

A circumscribed, rarefied and expanded lesion in a bone of a hand or a foot is likely to be an enchondroma. Calcific stippling makes the diagnosis a certainty. Actual cysts and giant cell tumors in this area are rare.

Prognosis and Treatment

In situations other than small bones malignant change is likely in about 25 per cent. This is particularly true when the pelvis is involved. The tumor should be excised or curetted and the wall cauterized. The capsule is removed to reduce the possibility of recurrence. A large defect is filled with bone grafts. Biopsy is avoided if possible, as the tumor is transplanted with ease. Recurrence is more likely in chondroma of the large long bones. When possible, removal from a finger should be avoided, as bone grafting is difficult. A sudden increase in size, persistent pain and spontaneous cortical perforation are ominous signs. Tumors of the large long bones and the pelvis should be looked upon with suspicion as potentially malignant, and radical resection should be done.

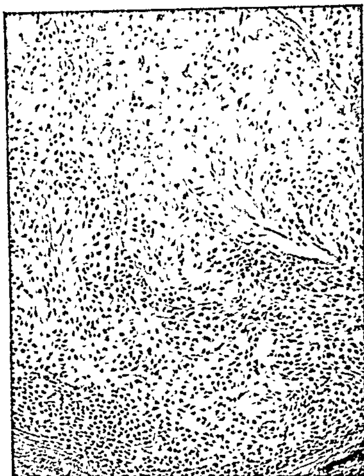
Periosteal Chondroma[8]

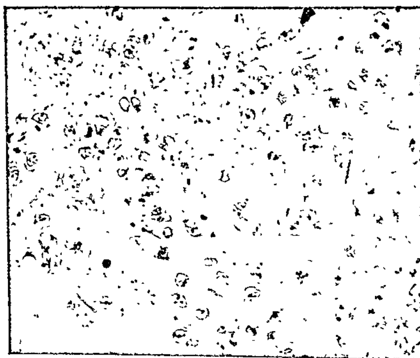
This benign cartilage tumor originates in periosteum, grows slowly, is small, and erodes *and induces sclerosis of contiguous cortical* bone. Roentgenograms show only a troughlike hollow in the sclerotic cortex. The patient complains of pain and gradual swelling. At operation a firm lobulated rubbery cartilage tumor adherent to the periosteum is found. The underlying cortex is gouged out and hard.

[7] Coley, B L., and Higinbotham, N L.: Significance of cartilage in abnormal locations, Cancer 2 777, 1949.

[8] Lichtenstein, L., and Hall, J. E. Periosteal chondroma, J Bone & Joint Surg. 34A·691, 1952.

PLATE 20. Chondrosarcoma. (*Top*, × 85) The typical transition from the embryonic tissue at the left to the wild-looking atypical cartilage at the right is well displayed. (*Bottom*, × 135) The features indicating malignancy include: pleomorphism, irregularity of arrangement, and plump, pyknotic, often multiple nuclei. The matrix characteristically stains poorly. Hypercellularity and presence of binucleate cells are important indications of malignancy.





Treatment consists of block excision and curettement of the bone.

CHONDROSARCOMA

Geschickter and Copeland include chondrosarcomas in the group of osteogenic sarcomas. Chondrosarcoma may be either primary or secondary, the latter arising from pre-existent cartilage tumors. However, certain writers, notably Phemister[9] and Jaffe and Lichten-

[9] Phemister, D B. Chondrosarcoma of bone, Surg, Gynec. & Obst. 50:216, 1930



FIG. 186 Chondrosarcoma of tibia This is the picture of a primary type of chondrosarcoma, but an antecedent cartilaginous exostosis is possible.

stein[10] stress the desirability of keeping chondrosarcoma apart from osteogenic sarcoma, because chondrosarcoma issues out of already formed cartilage, whereas osteogenic sarcoma arises from primitive osteogenic connective tissue. However, this author believes that the distinction is an arbitrary one. The following descriptions will be in keeping with the concept of chondrosarcoma arising as an alteration of the normal processes of osteogenesis, the process stopping short of ossification.

PRIMARY CHONDROSARCOMA
(Chondromyxosarcoma)

Primary chondrosarcoma is a very malignant tumor, composed of cartilage and myxomatous tissue and occurring subperiosteally, at first without involvement of the cortex.

Clinical Picture

AGE. 14 to 21 years

AREA OF PREDILECTION Knee, shoulder and pelvis

POSITION. At points of muscle attachment to bone near articular regions where cartilage formation persists throughout life; for example, where adductor magnus attaches to linea aspera.

SYMPTOMS. Pain becomes progressively more severe, and the knee undergoes flexion contracture. Pain is worse at night.

FINDINGS. Swelling of rubbery resiliency near a flexed knee joint.

COURSE. Very rapid. Fatal outcome.

Roentgenologic Findings. A dome-shaped translucent shadow is seen adjacent to but not involving the cortex or the medulla. Periosteal new bone is sparse but, like other subperiosteal tumors, may produce Codman's triangles at the corners. Mottling calcification shadows are scattered throughout.

Pathology. GROSS On removing the closely adherent periosteum, the tumor is composed of a lobulated, shiny, opalescent or semitranslucent substance. The cartilage contains white spots of calcification, cysts, hemorrhage and myxomatous tissue. Metastases occur by penetrating and growing within the lumen of regional veins. The lobules of cartilage are intimately connected to the surface of the bone, but the cortex is not penetrated except in advanced cases.[11]

MICROSCOPIC. Immediately beneath the periosteum, primitive embryoniclike connective tissue is found, composed of stellate or spindle-shaped cells. This tissue merges with a myxomatous syncytium which it forms. Beyond this the next step in cartilage formation is noted, consisting of formation of hyaline intercellular matrix which surrounds enscapulated cells with small dense nuclei and deeply acidophilic cytoplasm. Normally, the cells are arranged orderly in pairs or tetrads and tend to assume a columnar arrangement. Very subtle evidence of malignity should be sought in a frequently normal appearing cytologic picture. This consists of hypercellularity, plumpness of nuclei, and double nuclei. More pronounced evidence of malignancy is not common but is more likely to be found in recurrent tumors. These changes include: pronounced irregularity in size of the cells and their nuclei, the presence of numerous cells with multiple nu-

10 Lichtenstein, L, and Jaffe, H L Chondrosarcoma of bone, Am. J. Path 19 553, 1943

11 Coley, B L, and Higinbotham, N L Significance of cartilage in abnormal locations, Cancer 2 777, 1949.

clei, and hyperchromatism of the nuclei. Cell division tends to be amitotic, but a few mitoses may be seen. When the tumor is highly anaplastic, the cytologic process does not advance beyond the collagenous stage, and the picture of a fibrosarcoma is presented. Most frequently, the tumor tissue appears to be normal, and roentgenologic and clinical pictures must be correlated with the pathologic to arrive at a correct diagnosis.

Geschickter and Copeland stress the importance of the embryonic cellular tissue penetrating deeply within the tumor. This is in contrast with the benign tumor in which the primitive tissue remains peripheral. These observers interpret the myxomatous tissue as a phase in the formation of cartilage, whereas Kolodny and others consider it a product of degeneration.

Diagnosis. This tumor occurs in situations in which cartilaginous exostoses are prone to occur. It is not difficult to visualize a similar histogenesis for both types of growths. The periosteum retains its fetal chondrogenic potential throughout life. It is important to recognize that a very malignant growth can occur at this site. The characteristics denoting malignancy include: rapidity of growth, severe and persistent pain, and microscopic features such as hypercellularity, polymorphism, plump, multiple and dark nuclei and the presence of many multinuclear giant cells. Because cell division in chondrosarcoma tends to be amitotic, one should not seek mitotic division figures as evidence of malignancy.

Course. The tumor is not radiosensitive; it recurs locally and metastasizes with ease. A fatal outcome within a year is usual. The most radical form of amputation should be attempted.



Fig. 187. Chondrosarcoma. The location is common for enchondroma, which is revealed by calcified deposits within the shaft. A rapidly developing osteolytic destructive lesion eroding through the cortex, without surrounding reactive bone formation, accompanied by pain and appearing at the upper shaft of the humerus in an area of spotty calcification should suggest the diagnosis.

Secondary Chondrosarcoma

Secondary chondrosarcoma is a very slow-growing malignant tumor, occurring as a result of malignant degeneration of an enchondroma or a cartilaginous exostosis.

Clinical Picture

Age. Third decade or thereafter.

Site of Predilection. Long bones, especially the upper end of the humerus, the ribs and the innominate bone. Small bones of hands and feet are immune.

Position. Central chondroma is more predisposed.

Antecedent History. Insignificant, intermittent ache, swelling

Antecedent Lesion. Central chondroma of large bone, multiple hereditary cartilaginous exostoses, single exostosis, Paget's disease.

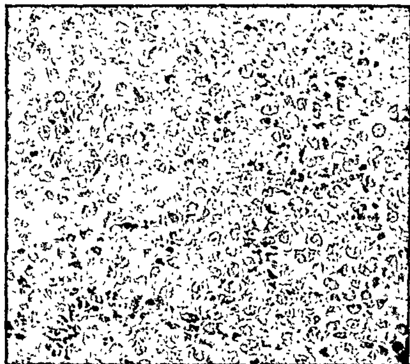


PLATE 21. Chondroblastoma. Large sheets of compact round and polyhedral cells with large vesicular nuclei. Marked vascularity. Giant cells. Transformation into cartilage not seen in this area. Reticulin fibers and calcium deposits are not demonstrable except by special staining, e.g., Rio Hortega stain. (× 310)

SUDDEN CHANGE OF SYMPTOMS. Pain and swelling increase, become more persistent, pain at night, edema due to venous and lymphatic obstruction

COURSE. Very slow, only locally invasive for a long time, 2 to 25 years

Roentgenologic Findings. The previous benign lesion presents evidence of malignancy by losing its sharp outline and becoming diffusely fuzzy. The main mass becomes stippled with many fragments of osseous tissue and focal areas of calcification The base or pedicle of the exostosis may persist as evidence of the original lesion. Chondrosarcoma arising in an exostosis generally remains external to the bone, and the cortex is little, if at all destroyed. In a central chondrosarcoma the outline of the tumor becomes vague, expanded, and mottled with calcific deposits The cortex becomes eroded very slowly and reacts by thickening considerably. The translucent shadow outside the bone usually does not calcify as extensively as the central portion. It may resemble myositis ossificans, but the latter rapidly forms longitudinal laminations of new bone.[12]

Pathology. Grossly, one may be able to identify the antecedent lesion, such as an exostosis, in the large cauliflower mass of cartilaginous tissue. The larger the tumor the more likely it is that it will display areas of degeneration such as cysts, necrosis, calcification and myxomatous tissue. The size or the appearance of the tumor is no indication of its malignancy. Even a normal-appearing cartilaginous exostosis or enchondroma should be suspected. Microscopically, evidence of malignity includes hypercellularity, plumpness of cells, polymorphism, plumpness of nuclei, which are often multiple and hyperchromatic, and reversion to the appearance of fetal cartilage throughout the tumor (large cells with scanty intercellular matrix). In a benign tumor a fetal type of cartilage is found only at the periphery, and formation of mature cartilage progresses in an orderly manner.

A central chondrosarcoma may be revealed only after removing what appears to be normal cortex. If the cortex has been eroded, it is thin and is penetrated with ease. The opening through which a central tumor penetrates to the exterior is often small compared with the large externally growing mass.

Treatment. Of all bone sarcomas, secondary chondrosarcomas offer the best prognosis Secondary chondrosarcomas are slow growing and, although incompletely removed, they may recur locally for a long time before invading and metastasizing through venous channels. The tumor may even grow within the vein over an extended period before spreading to

[12] Bloodgood, J. C. Bone tumors, myxoma, central and periosteal, Ann Surg. 72 712, 1920

the lungs. The degree of malignancy is greater in central chondrosarcomas. Treatment consists of amputation, which offers the highest incidence of cures. Irradiation is ineffective. Local removal almost surely will be followed by recurrence.

/CHONDROBLASTOMA

A chondroblastoma is a cellular, vascular and cartilaginous tumor of young adults, occurring about the epiphyseal line, destroying cancellous bone and characteristically containing multiple calcium deposits Jaffe and Lichtenstein and the majority of reports consider the tumor as benign.[13-18]

Clinical Picture

Age. Onset occurs before obliteration of the epiphyseal line, from ages of 10 to 20.

Sex. Males predominate

Area of Predilection. Ends of long bones about knee and upper humerus

Position. About one side of the epiphyseal line, chiefly in the metaphysis It may extend to the epiphysis.

Symptoms and Findings. Trauma, pain, tenderness, swelling in most cases. Occasionally, limp, joint effusion

Course. Rapid, from 1 month to 2 years

Roentgenologic Findings

Roentgenograms show a characteristic well-delineated area of rarefaction of cancellous bone extending over and beyond the epiphyseal line quite early. The main mass of tumor is either in the metaphysis or the epiphysis. The position is often eccentric. The cortex may be thinned but rarely penetrated. The tumor borders are irregular, fuzzy and vague.

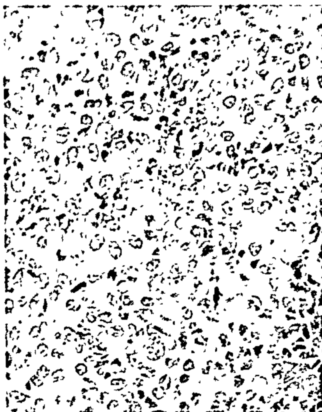


Fig. 188. Chondroblastoma. Microscopic appearance demonstrates sheets of closely packed cells with large vesicular nuclei.

This is in contrast with a giant cell tumor, which shows well-defined margins, and the cortex, although thinned, is elevated. Mottled areas of increased density throughout the tumor represent calcium deposits.

Pathology

Gross. At first glance the tumor appears as a dark-red, hemorrhagic and friable tissue within which are scattered small yellow zones of calcification, bluish-white translucent nodules of cartilage and a few spicules of necrotic bone. On closer inspection a soft grayish-pink tissue is discovered within the friable tissue representing the basic tumor tissue. When the tumor is less vascular and less necrotic, it is firm and grayish-pink and contains a variable amount of cartilage. White or yellow areas of calcium salts are found within the cartilage and in the tumor tissue. The tumor extends irregularly through the metaphysis, destroys the epiphyseal line at one side and occasionally invades the subperiosteal space. The necrosis and hemorrhage may suggest a giant cell tumor or malignant aneurysm of bone,

[13] Codman, E A · Epiphyseal chondromatous tumors of the upper end of the humerus, Surg, Gynec & Obst 52 543, 1931.

[14] Coley, B L, and Santora, A J Benign central cartilaginous tumor of bone, Surgery 22 411, 1947

[15] Copeland, M M, and Geschickter, C F Chondroblastic tumors of bone, benign and malignant, Am J Surg 129 724, 1949

[16] Jaffe, H L, and Lichtenstein, L Benign chondroblastoma of bone, Am J Path 18 969, 1942.

[17] Kolodny, A Bone sarcoma, Surg, Gynec. & Obst 44, 1927

[18] Valls, J, Ottolenghi, C E, and Schajowicz, F · Epiphyseal chondroblastoma of bone, J Bone & Joint Surg. 33A 997, 1951.



FIG 189. Giant cell tumor. Characteristics include loculated, osteolytic, expansile distention and thinning but not penetrating overlying cortex, epiphyseal location

but the chondromatous material, absence of new bone, and fusion with the epiphyseal line should clarify the diagnosis

Microscopic. Large sheets of compact round or polyhedral cells similar to mesenchymal tissue form the basic tumor tissue These cells are of moderate size and contain a relatively large nucleus, occasionally 2 nuclei Focal areas of calcification distributed throughout the cellular tumor tissue are typical A scattering of giant cells lies adjacent to areas of hemorrhage and necrosis. When calcification is unusually heavy, the tumor cells swell and undergo degeneration and necrosis, similar to cartilage tissue undergoing calcification preparatory to ossification, but osseous transformation does not occur. In some areas tumor cells can be seen undergoing transformation to cartilage cells. About necrotic zones, a reparative process takes place by resorption of calcific detritus and organization of hemorrhage.

The Rio Hortega stain will reveal calcium deposits and reticulin fibers about the tumor cells. Where the chondroblasts differentiate into cartilage, reticulum formation is rare. This indicates that the basic cell of this tumor is a reticulohistiocytic cell with a tendency to differentiate into chondroblastic tissue and then into cartilage.

Many blood spaces with and without a single layer of endothelium occur throughout the tumor.

TREATMENT

The majority of observers feel that this tumor is benign. Geschickter and Copeland believe that about 50 per cent become malignant. Cases have been reported where postoperative irradiation has been followed by malignant degeneration.

Conservative surgical treatment consists of curettage and obliteration of the cavity by bone grafts. In view of the possible untoward effects of irradiation, this form of therapy should be avoided.

BENIGN GIANT CELL TUMOR[19-22]

(Osteoclastoma)

This osteolytic tumor occurs in young adults at the epiphysis and is typified by an abundance of characteristic large giant cells.

CLINICAL PICTURE

Age. This tumor occurs in young adults after the epiphyseal plate has ossified and longitudinal bone growth is completed. It appears most commonly between the ages of 15 and 35.

Area of predilection is an asymmetric posi-

[19] Jaffe, H. L, Lichtenstein, L, and Portis, R B· Giant cell tumor of bone Its pathologic appearance, grading, supposed variants and treatment Arch. Path. 30 993, 1940

[20] Geschickter, C F, and Copeland, M M.: Tumors of Bone, Philadelphia, Lippincott, 1949.

[21] Bloodgood, J C: Benign giant cell tumor of bone; its diagnosis and conservative treatment, Am J. Surg. 37:105, 1923.

[22] Meyerding, H W· Treatment of benign giant cell tumor by resection or excision and bone grafting, J Bone & Joint Surg 27·196, 1945.

PLATE 22. Giant cell tumor. The tumor giant cell is very large and contains a tremendous number of centrally placed and uniform nuclei. The stroma is fibrous, vascular, and loose and contains spindle cells possessing large heavily chromatinized nuclei. Any tendency toward aggressiveness and malignancy should be sought in the fibroblasts which become more numerous, plumper, polymorphic, and contain darker nuclei and many mitoses. (× 240)



tion in the epiphysis of a long bone. The lower femur, the distal radius and the upper tibia are the common sites

Course. Chronic, a few months to several years

Sequence. Trauma, pain, tumor, pathologic fracture

Pain. Chronic, constant, in region of a joint, progressively more severe, worse at night, and increased with activity

Swelling. The end of the bone expands to one side, and overlying skin is stretched. In contrast with osteogenic sarcoma, the skin displays no dilated vessels. Pressure over the swelling produces audible and palpable crackling as the cortex is ruptured. Tenderness is moderate or absent.

Limitation of joint motion is not observed until late, as the tumor expands and distorts the end of the bone. There is no increase of joint fluid. The joint is not invaded.

Pathologic fracture occurs late in the course as a large extent of cancellous and cortical bone is destroyed

ROENTGENOLOGIC FINDINGS

The tumor is revealed by a large, sharply circumscribed area of reduced density asymmetrically located in the epiphysis, beginning [illegible] vard the me- [illegible] outward and [illegible] one formation is apparent. Multiple septae of bone and soft tissue traverse the interior and produce a characteristic loculated "soap bubble" appearance. As the tumor enlarges, the septae disappear. The cortex may be disrupted as the tumor invades and thickens the surrounding soft tissues. Extension into the joint is rare.

PATHOLOGY

Gross. The tumor is composed of ragged, very friable, readily bleeding tissue containing variously sized cavitations and small cysts. These cavities may be filled with old or fresh blood, detritus of degenerated tissue, or a mucinouslike material. The color of the tumor varies from a reddish or chocolate brown, in which the vascular tissue predominates, to grayish or mottled, wherein the connective tissue is the major component. The epiphyseal end of the bone is distorted and enlarged to one side. It is invested by periosteum and cortex, which is thinned and fragile, being easily broken by handling. The underlying bloody tissue imparts a blue color through the transparent thin cortex. No periosteal new bone formation can be discerned grossly. The inner wall of the tumor is lined by a fibrous capsule from which septae extend inward to partition the tumor.

Microscopic. At the periphery, a fibrous capsule envelops the tumor. Beyond this, a thin layer of new imperfectly formed bone is

seen. The capsule of fibrous tissue and new thin bone explain why the tumor is sharply demarcated and expands rather than infiltrates. The characteristic finding in the tumor tissue is the presence of an abundance of tumor giant cells. These large cells measure from 10 to 100 microns and contain many centrally placed uniformly sized nuclei. The number of nuclei varies from 15 upward, some estimates being as high as 150. The giant cells are numerous about areas of hemorrhage, spicules of old bone, and walls of small cysts. By comparison, the Langhans giant cells characteristic of tuberculosis contain a smaller number of peripherally placed nuclei; foreign body giant cells are smaller, contain a lesser number, usually less than 15, of variously sized, centrally placed, nuclei. The stroma consists of a vascularized loose network of spindle-shaped and ovoid cells heavily interspersed with tumor giant cells. The spindle cells are oval, elongate and contain a relatively large, heavy chromatinized, nucleus and a small amount of acidophilic cytoplasm Fibrils about these cells suggest their fibroblastic function. The appearance of the spindle cells indicates the malignant potential of the tumor. The tumor aggressiveness is proportionate to the increase and the crowding of the stromal cells and the plumping up of these cells, the increase of chromatin in the nuclei, variation in the sizes and the shapes, and the presence of many mitotic figures. The more the atypism, the greater the tendency to metastasize. A benign giant cell tumor displays uniformity of cells. At the periphery of the tumor, spicules of old ragged bone with giant cells closely applied to their surface are seen.

Differential Diagnosis

Giant cells are frequently found in other lesions, the so-called variants of giant cell tumors. These lesions include nonossifying fibroma, unicameral bone cyst, localized osteitis fibrosa, aneurysmal bone cyst, chondromyxoid fibroma, benign chondroblastoma, and the "brown tumors" sometimes found in hyperparathyroidism. The following characteristics should be sought in identifying a giant cell tumor.[23]

1. The patients are nearly all young adults.
2. The majority of giant cell tumors originate in the end of long bones in the portion which during the growth years was the epiphysis.
3. Pain, often present at night as well as during activity, is common. By contrast, pain is almost never associated with a solitary bone cyst, fibrous dysplasia, nonossifying fibroma, etc.
4. Tumefaction is present early and is palpable and tender.
5. The expanded cortex is paper-thin and may crackle when palpated.
6. Roentgenograms show a circumscribed osteolytic but expansile tumor, and not uncommonly the thin expanded cortex of bone is perforated. The larger giant cell tumors may appear multilocular and resemble soap bubbles.
7. Periosteal new bone formation is not apparent on roentgenograms.
8. The stromal cells in a giant cell tumor show greater growth and multiplication and variety of morphology than do the stromal cells of other lesions which commonly contain giant cells.
9. Stromal and giant cells in a giant cell tumor contain acid phosphatase but no alkaline phosphatase.
10. In other lesions, giant cells are fewer in number. The stromal cells of lesions which are not true giant cell tumors contain only alkaline phosphatase and never acid phosphatase.

Treatment

The limb is conserved where possible. The tumor is curetted, and the wall is thoroughly cauterized. The cavity is filled with bone chips. When the tumor is extensive and the retaining cortical walls are deficient, resection of that segment of the bone is done, and the remaining shaft is arthrodesed to the other bone at the joint. An intramedullary pin is an effective way of accomplishing this.

Irradiation should be avoided; frequently, it is of no benefit and may produce extensive soft tissue damage and cause the tumor to become malignant. Likewise, radium implantation into the cavity postoperatively should be avoided, as it may cause intractible infection which necessitates amputation. Recurrence is characteristic of these tumors, particularly

[23] Compere, E L. The diagnosis and treatment of giant-cell tumors of bone, J Bone & Joint Surg 35A·822, 1953.

PLATE 23. Aneurysmal bone cyst. Characteristic blood-filled lakes are surrounded by reactive bone which forms by metaplasia of the fibrous stroma. (× 30)



when the wall is not cauterized Postoperative irradiation does not seem to prevent recurrence and may even cause osteogenic sarcoma However, this treatment may be used in inaccessible places, such as the skull and the vertebrae.

In the event of recurrence, reoperation is indicated, and microscopic examination for malignant changes is conducted at that time. Resection is advisable if the lesion is in the fibula, the radius, the ulna or the humerus. Amputation is indicated for malignant giant cell tumor.

SUBPERIOSTEAL GIANT CELL TUMOR

This is a peculiar variant characterized by rapid development after trauma of an osteolytic lesion containing blood. The tissue consists of giant cells in an ossifying stroma of fibrous tissue No expansion of cortex occurs. Reossification and healing are spontaneous. This tumor is described under "Bone Aneurysm."

ANEURYSMAL BONE CYST
(Ossifying Subperiosteal Hematoma, Subperiosteal Giant Cell Tumor)

This benign bone lesion consists of a mass of vascular spaces enclosed in a shell of periosteal n[illegible] and dis[illegible]

CLINICAL PICTURE

Age. Occurs most commonly between 10 and 30.

Sex. Males predominate.

Area of Predilection. The metaphyseal region of long bones. Less often, flat bones and vertebrae. In a vertebra, the arch is usually affected; the body, infrequently.

Symptoms and Findings. A history of trauma often antedates pain accentuated by movement, bony swelling and limitation of joint motion. When a vertebra is involved, signs of spinal cord or nerve root pressure are found.

ROENTGENOLOGIC FINDINGS

The affected bone is expanded, cystic and ballooned outward. The mass is roughly ovoid, displays a slightly increased soft tissue density, extends outward eccentrically from the bone, destroys the original cortex and is surrounded by a faint outline representing periosteal new bone. The radiolucent tumor gradually becomes mottled and coarsely trabecu-

[24] Lichtenstein, L · Aneurysmal bone cyst, Cancer 3·279, 1950

[25] Thompson, P. C. Subperiosteal giant cell tumor, J. Bone & Joint Surg. 36A:281, 1954.



Fig. 190. Aneurysmal bone cyst of the cervical spine. The roentgenogram reveals the soft tissue mass (hemorrhage) along the anterior aspect of the 3rd, the 4th and the 5th cervical vertebrae and the osteolytic lesion of origin in the antero-inferior angle of the body of the 4th cervical vertebra. The mass is just beginning to ossify. Clinically, the onset is sudden and produces pain, torticollis, restriction of neck motion, and dysphagia. Examination reveals a tender, fixed, ligneous mass which is not visible in roentgenograms until ossification develops.

lated and eventually may completely ossify. Although the underlying cortex is destroyed and the medullary canal exposed, the shaft is not expanded. When the tumor involves a vertebra, the arch is usually affected. However, when situated within the vertebral body, it must be differentiated from a hemangioma. The vertical striations peculiar to hemangioma are never observed in an aneurysmal bone cyst.

Pathology

Gross Pathology. The large mass is attached



Fig. 191. Aneurysmal bone cyst. The walls of connective tissue display new bone formation and giant cells which are smaller than the tumor type of giant cells.

by a broad base to the shaft of a long bone growing outward and displacing the soft tissues. It is surrounded by a thin shell of bone enclosing cystic blood-filled spaces. The thin bony shell is easily penetrated, and a reddish-brown, liverlike friable mass interspersed with gritty particles of bone is encountered. A semiliquid substance having the appearance of partially organized blood clot lies in the center. Occasionally, the soft center may be continuous with the medullary canal. The highly vascular tumor bleeds profusely but slowly on attempted removal. Fibro-osseous septae extend throughout the tumor.

Microscopic Appearance. The spongy bone and marrow are replaced by small and large pools of blood enclosed in fibro-osseous septae. The supportive connective tissue bordering the vascular spaces contains multinuclear giant cells, new bone formation and calcium deposits. The giant cells are small and contain small nuclei in contrast with large tumor giant cells. The peripheral shell displays active periosteal new bone formation.

Treatment

These lesions may regress spontaneously and ossify. X-ray therapy hastens the process. Before the tumor becomes extensive, it is advisable to curette the lesion, fill the defect with bone grafts, and follow with x-ray therapy.

UNICAMERAL BONE CYST

This lesion usually occurs in the first two decades of life, particularly between the ages of 9 and 14.[26] The long bones are almost invariably involved, but the calcaneus is often the site. Most lesions are found at the proximal end of the humerus or the femur. Because *the cyst occurs in an area of growth and bone remodeling at an age when these processes are active*, disturbance of such processes seems to be the most tenable theory of origin. Clinically, the only symptom is a mild ache or pain in the affected area. The typical roentgenographic features are: (1) destruction of medullary bone; (2) destruction of bone on the inner side of the cortex; (3) subperiosteal



FIG. 192. Unicameral bone cyst with pathologic fracture.

new-bone formation, giving the appearance of expansion; (4) an expansile defect in the metaphysis; (5) trabeculated lines in the wall of the cyst, giving the appearance of multiloculation; (6) when the lesion is old, a thin, dense cyst wall is evident; (7) serial films show the *lesion gradually displaced away from the epiphysis*. Jaffe and Lichtenstein regard the cyst as actively enlarging while it is adjacent to the epiphyseal plate, and "latent" when it is distal and separated from the epiphyseal plate by an area of normal trabeculated bone.

Grossly, the bone displays an area of fusiform expansion. The periosteum lifts away easily, and the underlying bone is thin and bluish. Penetration of the thin cortex is easy. The cavity contains yellow fluid. Trauma may cause the fluid to be hemorrhagic. A thin layer of connective tissue lines the inner surface of the wall, which displays multiple scroll-like elevations. The cyst wall of bone is thin. Microscopically, nothing characteristic is seen.

Differential diagnosis includes fibrous dysplasia [illegible]
encho
from
lipoma.

The most satisfactory treatment is excision combined with bone grafting. Recurrences are less likely by this procedure, but when they occur are usually in individuals below the age of 10 and when the lesion is juxta-epiphyseal, the so-called "active" type.

Pathologic fracture may occur in as high as 50 per cent of cases, with hemorrhage into the cavity and spontaneous healing and reossification.

[26] Garceau, J. G., and Gregory, C. F.: Solitary unicameral bone cyst, J. Bone & Joint Surg. 36A:267, 1954.

OSTEOGENIC SARCOMA

This malignant tumor of bone is derived from bone-forming elements and is characterized by the presence of any of the tissues formed in the normal process of bone formation, namely, osteoblasts, osteoid, bone and cartilage.

SCLEROSING FORM

Clinical Picture

ONSET. Trauma frequently initiates symptoms.

SYMPTOMS. Pain, particularly at night, antedates all symptoms and findings.

FINDINGS. Swelling of bony hardness develops near the end of a long bone. Soft parts over the tumor gradually become immobile. Overlying skin may become thin and glossy but never ulcerates. Absence of inflammatory signs. Systemic reaction as fever and leukocytosis is rare but when present is low grade.

COURSE. Progressive over a period usually less than 10 months.

PREDOMINANT LOCATION. Lower femur and upper tibia. Less commonly the upper humerus, small bones of the hands and the feet and the vertebrae.

AGE. Young adults 15 to 25 years.

Pathology

GROSS. The tumor develops in the metaphysis. The epiphysis is never involved primarily, the epiphyseal plate walling off the tumor within the shaft, and the attachment of the periosteum at the epiphyseal line restraining the tumor in the subperiosteal area. The tumor is a white or gray tissue whose density varies between that of fibrous tissue and bone It may appear to develop mainly in the subperiosteal space and infiltrates toward the medulla through the cortex, or the main mass may be within the medulla, occupying the angle between the epiphyseal plate and the cortex and infiltrating outward through the cortex into the subperiosteal space. Within the tumor mass new bone formation is visible as grains which generally run outward at right angles to the shaft The bony substance imparts a gritty sensation to the touch. Eventually, the growing tumor penetrates the periosteum and invades the surrounding soft tissues. The oldest portion of the tumor nearer the cortex is more compact. As the periosteum is elevated from the surface of the shaft, it excites new bone formation in an effort to wall off the invading tissue. This new bone is seen best at the upper and the lower angles of the subperiosteal space. Within the shaft, the neoplasm extends distally and, after disappearance of the epiphyseal plate, proceeds into the epiphysis. The joint is practically never invaded. Metastases occur through the blood stream to the lungs.

Microscopic. Basically, this consists of proliferation of connective tissue, spindle cells, osteoblasts, osteoid and new bone. The osteoblasts are abundant, large, and contain large, dark-staining, vesicular nuclei. The contour of the cell, although typically polyhedral, is mainly pointed in one direction. Between the cells is found abundant deep-staining osteoid arranged in disorderly fashion. The osteoblasts exhibit anisocytosis, poikilocytosis, mitotic figures, and are not arranged in orderly rows about bone spicules. Rarely, an admixture of cartilage and a mesenchymal type of tissue are found.

Roentgenologic Findings. The basic picture is one of increased density due to increased bone formation.

EARLY. The cortex and the marrow appear to be undisturbed. Fine lines of increased density radiate laterally from and at right angles to the surface of the shaft, giving the typical "sun-ray" appearance. Or, a wedge of dense bone is found beneath the cortex and behind the epiphyseal plate.

LATER. The increased density of new bone permeates throughout the metaphysis. The periosteal bone is dense, and the cortex and the marrow cavity are invaded and appear sclerosed The markings of normal bony architecture become obliterated, particularly in the characteristic triangle between the epiphyseal plate and the cortex. Some secondary destruction produces a mottling effect. As the periosteum is raised, a triangle of ossification forms at the upper and the lower angles (Codman's triangle). This is not characteristic of neoplasm alone. Any process, as infection, which raises the periosteum produces this reactive bone.

Prognosis. The duration is generally less than 10 months. Following radical amputation,

a 5-year survival can be expected in about 20 per cent of cases. If no recurrence is demonstrable 18 months after surgery, permanent cure is probable.

Treatment. Radical amputation offers the only hope of cure. Inoperable types may secure palliation by implantation of 50-mg. estradiol pellets and administering stilbestrol and dicalcium phosphate with vitamin D by mouth. These tumors are not radiosensitive.

Experimentally Induced Osteogenic Sarcoma.[27] Osteogenic sarcoma can be produced in rabbits by intravenous injection of beryllium compounds. Beryllium, a rare element, is used industrially as a constituent of alloys, and its fluorescent property is useful in fluorescent lamps. The radioisotope Be^7 is deposited chiefly in the skeleton, especially the long bones. Beryllium is the most toxic of all the elements, and symptoms of berylliosis can develop even after inhaling the smallest amounts. It is excreted very slowly through the kidneys. When osteogenic sarcoma develops, splenic atrophy is an invariable accompaniment. This suggests the possible use of splenic substances to determine its effect on these tumors.

Secondary Osteogenic Sarcoma. When this tumor develops at an age after the growth period, it generally arises secondary to benign lesions such as the callus of fracture healing, myositis ossificans, fibrous dysplasia, Paget's disease and multiple cartilaginous exostoses.

Irradiation Sarcoma.[28, 29] The only definitely established causes of sarcoma of bone in man are roentgen irradiation and radioactive elements. Dial painters repeatedly ingest minute amounts of radium, mesothorium and radiothorium and demonstrate radioactivity in all the bones Sarcoma is a definite occupational hazard in these individuals as well as in workers dealing with atomic energy development.

[27] Janes, J M, Higgins, G. M, and Herrick, J. F · Beryllium-induced osteogenic sarcoma in rabbits, J. Bone & Joint Surg 36B·543, 1955.

[28] Hatcher, C H Development of sarcoma in bone subjected to roentgen or radium irradiation, J Bone & Joint Surg 27 179, 1945

[29] Martland, H S A general review of data gathered in the study of the radium dial painters with special reference to the occurrence of osteogenic sarcoma, Am J Cancer 15 2435, 1931.

Osteolytic Form
(Malignant Bone Aneurysm; Telangiectatic Sarcoma)

This is a destructive tumor which arises in the marrow cavity of long bones. It occurs over a wider age distribution than the sclerosing form, although it is most common in young adults. It possesses an unusual tendency to pathologic fracture which does not heal.

Clinical Picture

Age. All ages but commonest at 10 to 20.

Predominant Location. Long bones, especially at lower femur and upper tibia. It usually starts in metaphysis in the medulla and spreads rapidly to the epiphysis and the rest of the shaft.

Pathologic Fracture Frequent. Occurs in 50 per cent, failure of union.

Symptom Sequence. Pain, tumor, limp, trauma, fracture

Findings. A peculiar boggy swelling with suggestive fluctuation is apparent as the soft tissue is invaded. The overlying skin is tense, not inflamed, and contains dilated veins. Pulsation may be present and suggests an aneurysm. Invasion of the epiphysis and the surrounding soft tissues results in limitation of joint motion. Abnormal mobility and crepitus indicate a fracture.

Course. Progressive over a period of 2 years. In a small percentage, the course is chronic over several years.

Pathology

Gross. This is a vascular tumor which resembles a blood clot or recent active hemorrhage. It is a soft, friable, bloody tissue interspersed with fibrous tissue which encloses secondary hemorrhagic cysts. The main mass of the tissue is found subcortically, although the cortex is moth-eaten, and penetrated so that the tumor comes to lie subperiosteally; finally, the surrounding soft tissues are infiltrated and present a ragged bloody appearance. The predominant location of the tumor is in the metaphysis and extends distally in the marrow cavity of the shaft and proximally into the epiphysis. The bone usually fractures transversely at the metaphysis.

Microscopic. The picture is one of blood vessel spaces without an endothelial lining lying in tissue composed mainly of plump spindle cells and round abortive osteoblasts

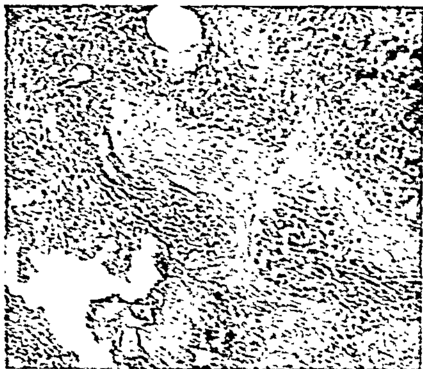


PLATE 24. Osteogenic sarcoma. Note the marked cellularity, pleomorphism, mitoses, poorly organized trabeculations and hypercellular disorganized cartilage. (× 140)

displaying extreme anaplasia, hyperchromatism and abundant mitoses. A variable amount of osteoid is scattered throughout, but no actual osseous tissue is formed. Giant cells are present in varying degree. The foreign body type is common in the slow-growing tumors. The rapidly growing more malignant growths display the multinucleated tumor giant cells which may confuse the diagnosis with that of a malignant giant cell tumor. Therefore, the characteristic findings are:

1. Plump spindle cells with dusty nuclei
2. Large round cells with numerous mitoses classified as osteoblasts
3. Giant cells are sparse and small and contain less than 15 nuclei.
4. Osteoid tissue, scattered sparsely. Osseous tissue, rare or absent.



FIG. 193. Osteogenic sarcoma, roentgenographic appearance, anteroposterior view. At the epiphysis and metaphysis of the femur, note: (1) loss of trabeculations, (2) mottled effect of destruction and sclerosis, (3) "sun-ray" extending from medial aspect of epiphysis.

The preponderance of malignant spindle cells requires differentiation from fibrosarcoma. In the latter, whorl formation is found, osteoblasts are rare, and osteoid is absent. It is also distinguished from the sclerosing form by the extreme anaplasia of osteoblasts which lie free in a stroma of spindle cells rather than about bone spicules and by the scarcity of osseous tissue. It is differentiated from chondromatous sarcoma by the lack of cartilage.

Roentgenologic Findings. The typical finding is a central area of irregular destruction extending through an unexpanded cortex resulting in periosteal reaction. The osteolytic area at first is subcortical and metaphyseal. The cortex is gradually thinned, and as the periosteum is elevated, the rays of reactive bone develop at right angles to the shaft. However, no reactive bone surrounds the

Fig. 194. Osteogenic sarcoma, roentgenographic appearance. (*Left*) Lateral view. Destruction, sclerosis and mottled appearance. The location is typical. (*Right*) Roentgenogram of specimen from which soft tissue has been removed. Subperiosteal extension of the tumor and radiating rays of new bone are amply demonstrated.

tumor anywhere. Later, the bone appears extremely moth-eaten, and a transverse ununited fracture at the metaphysis may be apparent.

Treatment. The chronic slow-growing tumor offers the greatest hope of cure, especially where it seems to arise secondarily from some pre-existent pathology as Paget's disease in the adult. Radical amputation may obtain a 5-year cure in as high as 20 per cent. The tumor is sensitive to irradiation. Inoperable cases should be irradiated, and calcium and estrogenic therapy instituted.

Summary on Osteogenic Sarcoma

Osteogenic sarcoma is a rapidly malignant tumor with a characteristic of producing osseous tissue. It is important to identify this bone-growing potential which infers a serious outlook. The tumor may be histologically similar to fibrosarcoma or chondrosarcoma, which have a distinctly less malignant prognosis.

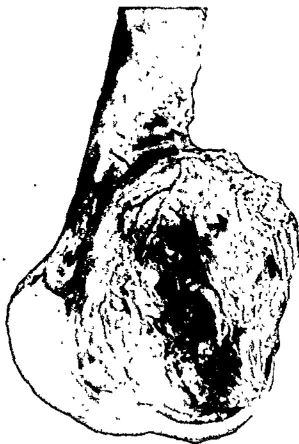


Fig. 195. Osteogenic sarcoma, gross specimen.



Fig. 196. Osteogenic sarcoma. Demonstrating the characteristic "sun-ray" appearance.

The tumor contains any one or all the elements of osteogenesis, including osteoblasts, osteoid, bone and calcifying cartilage. The finding of only one of these components is sufficient to classify this as an osteogenic sarcoma. Fibroblasts are likewise subject to anaplastic changes in this neoplasm, inasmuch as they stem from a common origin.

Osteogenic sarcoma varies in appearance, depending on the predominating component. At one extreme, the neoplasm is soft and highly vascular, embryonic mesenchymelike tissue is abundant, and a profusion of anaplastic polyhedral osteoblasts occupies the main mass. Anaplasia of osteoblasts is evidenced by their large size, nuclei which are large and hyperchromatic, and frequent mitoses. The process seldom proceeds to the

Fig. 197. Problem: Is this a chondrosarcoma or an osteogenic sarcoma? By Geschickter and Copeland's classification, because of the presence of neoplastic new bone it is called the *chondrosarcoma type of osteogenic sarcoma.* However, true osteogenic sarcoma develops new bone in the sarcomatous stroma and carries a grave prognosis. By Lichtenstein's classification, because the new bone is developing by metaplasia from cartilage, a potential of any cartilaginous tissue, the tumor is rightfully termed a *chondrosarcoma*. This has a lesser degree of malignity.





PLATE 25. Osteoid osteoma. Abundant pink-staining osteoid tissue surrounded by a vascular mesenchymal stroma containing proliferating fibroblasts and osteoblasts. (× 110)



point of mature bone formation, so that several areas must be examined before evidence of osteogenesis is found. This soft vascular tumor constitutes the osteolytic type of osteogenic sarcoma.

At the other extreme, the tumor is strongly osteogenic. Anaplastic osteoblasts form in irregular fashion osteoid and bone which become intermingled with unresorbed normal bone trabeculae.

Between the two extremes lie the more common type, which displays a moderate amount of osteogenesis revealed roentgenographically as increased density, obliterating lines of normal bone trabeculation.

Rarely, cartilage may form the major component, but evidence of calcification and transformation to osseous tissue distinguishes this as osteogenic sarcoma by Geschickter and Copeland's classification. Lichtenstein and others insist on formation of neoplastic bone by the sarcomatous stroma before the tumor can be called an osteogenic sarcoma. Cartilage always has the potential for forming bone by metaplasia, so that when this type of ossification is exhibited the tumor should be classified as a chondrosarcoma, which has a lesser degree of malignity than an osteogenic sarcoma.

When fibroblastic elements form the main mass of the tumor, the presence of neoplastic new bone distinguishes this from true fibrosarcoma.

Roentgenographic recognition of an osteolytic type of osteogenic sarcoma is difficult. Serial studies may eventually reveal mottled foci of osteogenesis, but one should not await this finding before attempting a biopsy.

OSTEOID OSTEOMA

Jaffe[30] was the first to give an accurate description and establish this tumor as a definite entity. However, it was Bergstrand[31] in 1930 who published the first description of the pathology. Defined, it is a small rarefying lesion in enchondral bones, composed of vascular fibrous tissue, proliferating fibroblasts and minute spicules of newly formed osteoid.

CLINICAL PICTURE

Young adults, especially males, from 10 to 25 are predisposed, although the condition occurs from 5 to 35. It has a predilection for the long bones, particularly the tibia and the femur. The small bones of the hands and the feet may be involved. It is a solitary lesion which causes pain, mild at first, then becoming

[30] Jaffe, H. L · Osteoid osteoma: a benign osteoblastic tumor composed of osteoid and atypical bone, Arch Surg 31:709, 1935.

[31] Bergstrand, H · Über eine eigenartige, warscheinlich bisher nicht beschreibene osteoblastiche Krankheit in den langen Knochen der Hand und des Fusses, Acta radiol. 11:597, 1930.

progressively more severe, continuous, agonizing, and worse at night The individual may limp. Gradually, a localized swelling which is palpable as a bony enlargement fusiform in shape becomes manifest. It is tender, sometimes exquisitely so. When the lesion occurs in the spine, acute localized back pain, muscle spasm, secondary scoliosis and pelvic tilt form the clinical picture. Systemic symptoms are absent. The temperature is normal, and blood counts are negative. The overlying skin is not reddened or warm.

Roentgenologic Findings

A solitary small rarefied lesion, usually less than 2 cm. in diameter, is found in the cortex, the subcortical region or the subperiosteal area, surrounded by thickened and sclerotic bone. The surrounding reactive bone may attain large proportions and is fusiform in shape It may even obscure the nidus. Occasionally, a small dense center of ossification may be seen in the nidus. When sciatica is the presenting complaint, the femur should be x-rayed routinely. An incidence of as high as 5 per cent of osteoid osteoma lesions may be found.

Microscopic Appearance[32, 33]

A very vascular mesenchymal type of connective tissue stroma with proliferating fibroblasts and osteoblasts surrounds abundant pink-staining osteoid tissue. The osteoid forms an irregular branching network. At the center of the lesion, a minute amount of poorly formed osseous tissue sometimes forms, but this may be entirely lacking. Surrounding this tissue is marked bony proliferation which is more marked when the lesion is intracortical. The marrow is somewhat fibrous. No leukocytes can be found, suggesting the noninflammatory nature of the lesion. Cartilage is never found.

Differential Diagnosis

The condition should be differentiated from:

1. Sclerosing nonsuppurative osteomyelitis of Garré
2. Benign bone cyst
3. Brodie's abscess
4. Chronic osteomyelitis with annular sequestrum
5. Syphilitic ossifying periostitis
6. Eosinophilic granuloma

Treatment

Complete excision of the nidus immediately and effectively relieves the pain. It is not necessary to remove the surrounding sclerotic bone, as this recedes after removal of the center.

PAROSTEAL OSTEOMA

Parosteal osteoma is a tumor of heterotopic ossification in a fibrous stroma similar to myositis ossificans but has a more intimate relationship to bone and a graver prognosis Benign and malignant forms occur, the latter predominating.[34]

Initial Lesion

This consists of a benign proliferation of ossifying fibrous tissue, which results in a rounded bony mass projecting from the shaft of a long bone at or near the metaphyseal region. It does not form a pedicle of normal orderly bone or a cartilaginous cap and in no way is related to an exostosis. The ossifying mass eventually invades the adjacent cortical and cancellous structure and extends outward peripherally into the soft parts with or without a periosteal enscapulating membrane. Islands of cartilage may be found within the ossifying mass. At the periphery, minute areas of malignant-appearing spindle cells or osteoblasts may be found. The tendency is toward progressive growth and ultimate malignant change, the histologic picture resembling sclerosing osteogenic sarcoma or spindle cell sarcoma. The area of predilection is over the surface of long bones which are free of muscular attachment. A common site is the floor of the popliteal fossa. Individuals between 20 and 40 seem to be predisposed.

Clinical Findings

A mass of bony hardness, which is tender, is fixed to the subjacent bone. Moderate pain

[32] Geschickter, C F., and Copeland, M M Tumors of Bone, Philadelphia, Lippincott, 1949.

[33] Sherman, M S · Osteoid osteoma, J Bone & Surg 29.918, 1947

[34] Geschickter, C. F, and Copeland, M. M · Parosteal osteoma of bone: a new entity, Ann. Surg 133·790, 1951.

and limitation of motion in the adjacent joint may be complained of.

Roentgenologic Findings

Roentgenograms reveal a dense irregular or rounded mass of new bone separated from the underlying skeletal structure. The underlying cortex may appear eroded an flattened but also displays sclerosis. At operation, a hard bony mass is encountered in the soft tissue, fused with the periosteum but encapsulated.

Grossly, cut sections of the mass display a firm, dry, striated surface simulating fibrosed cancellous bone. Microscopically, adult bone and fibrous tissue predominate. Foci of fibrospindle cell sarcoma are identified at the periphery. The osseous spicules are embedded in a vascular connective tissue. At a late stage, evidence of malignancy appears throughout the sections, imitating sclerosing osteogenic sarcoma or fibrospindle cell sarcoma.

After surgical removal, irradiation to the area is indicated. Any suspicion of malignancy should be cause for amputation. After local resection, the masses recur, spread through the soft tissues and penetrate the bone. Death results from pulmonary metastases.

The differential diagnosis lies between localized myositis ossificans, intraligamentous osteoma, osteoma of a tendon sheath, and sclerosing osteogenic sarcoma.



Fig. 198. Parosteal osteoma. Roentgenographic appearance. (*Left and center*) Lower femur. (*Right*) Upper end of ulna, left arm. (Geschickter, C. F., and Copeland, M. M.: Parosteal osteoma of bone, Ann. Surg. 133:790)

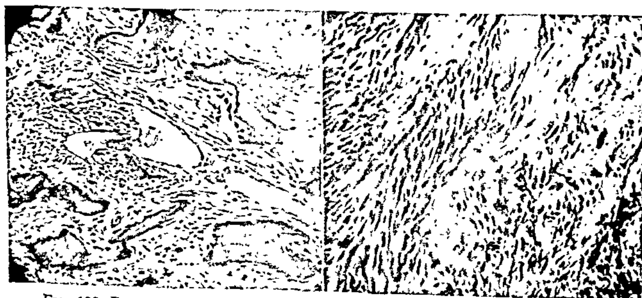


Fig. 199. Parosteal osteoma. (*Left*) Shows normal bone spicules forming in fibrous tissue. (*Right*) Typical fibrospindle sarcoma. (Geschickter, C. F., and Copeland, M. M.: Parosteal osteoma of bone, Ann. Surg. 133:790)



PLATE 26. Ewing's tumor. The sheets of cells tend to be grouped in lobules. A characteristic pseudorosette formation is well displayed. The bone is necrotic in some areas because vessels are choked off by tumor cells. (× 95)



PLATE 27. Ewing's tumor, higher magnification. The tumor consists of small polyhedral cells with ill-defined borders, meager cytoplasm, and nuclei which are large, uniform, prominent, round or oval and contain powdery blue-staining chromatin. The nucleus is more than twice as large as that of a lymphocyte. No interstitial stroma is seen. The dark-staining ring-shaped accumulation of cells envelops a center of necrotic cells and therefore is termed a "pseudorosette" in contradistinction to the rosette of a neuroblastoma whose center contains neurofibrils. (× 240)

EWING'S SARCOMA[35-41]
(Undifferentiated Round Cell Sarcoma; Endothelial Myeloma)

Theoretically, two types of tumors may arise from the bone marrow. From the marrow cells themselves arises the myeloma. From the supportive marrow structure composed of reticuloendothelial tissue develops Ewing's tumor, reticulum cell sarcoma and hemangio-

[35] Campbell, W C, and Hamilton, J F Gradation of Ewing's tumor, J Bone & Joint Surg 23 869, 1941.

[36] Coley, W. B.: Endothelial myeloma or Ewing's sarcoma, Am J. Surg 27·7, 1935.

[37] Ewing, J. Endothelial myeloma of bone, Proc New York Path Soc. 24 93, 1924

[38] Geschickter, C. F, Copeland, M. M, and Maseritz, I. H: Ewing's sarcoma, J. Bone & Joint Surg 21 26, 1939

[39] Jaffe, H L Pathology, problem of Ewing's sarcoma, Bull Hosp Joint Dis 6 82, 1945.

[40] Kolodny, A· Bone sarcoma, Surg, Gynec & Obst. 44 126, 1927.

[41] Willis, R A Metastatic neuroblastoma of bone presenting the Ewing's syndrome, with a discussion of Ewing's sarcoma. Am J. Path 16:317, 1940.

endothelioma. The majority of observers believe that the reticulum cell is the stem cell for Ewing's tumor and reticulum cell sarcoma. Others favor a lymphogenous (Geschickter and Copeland) or a sympathetic tissue origin (Willis).

Ewing's tumor is a malignant tumor typified by its occurrence in childhood, clinical findings of fever and leukocytosis, a characteristic onion-peel periosteal shadow in roentgenograms, and extreme radiosensitivity. A fatal outcome is invariable.

Clinical Picture

Age. 4 to 25. Caucasians almost exclusively

Area of Predilection. The long bones, although other bones may be affected.

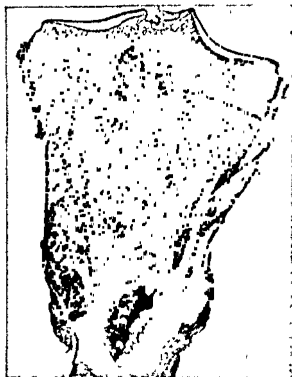


Plate 28. Ewing's tumor of tibia. The necrotic tissue in the diaphysis represents the remains of the tumor after irradiation. Freshly removed tumor tissue which has not been exposed to x-rays will stain blue. Note elevation of periosteum by reactive new bone.

Fig. 200. Ewing's sarcoma. Typical "onion-peel" appearance.

Location. The diaphysis

Symptoms. *Pain,* intermittent, recurring with greater intensity and persistency, worse at night. *Tumor,* palpable, indurated, tender, and fixed to underlying bone; skin reddened, edematous and contains dilated veins. *Constitutional reaction,* including fever, leukocytosis and anemia.

Course. Periods of *remissions* with decrease in size of the tumor, and *exacerbations* with increased size of the tumor, over a period of months to a few years. *Metastasizes to other bones* particularly the skull, the vertebrae and the ribs. *Metastasizes late to the lungs,* causing chest pain and hemoptysis. Metastatic spread takes place through both lymphatics and blood stream.

Roentgenologic Findings

At the midshaft of a long bone, the cortex displays at first increased density, which extends externally as periosteal new bone forming multiple thin layers parallel with the surface of the shaft. This is called the "onion-peel" appearance, most frequently found in association with this tumor, but occasionally this type of reactive bone formation develops in response to elevation of the periosteum by other tumors and subperiosteal infection. In addition, some of the periosteal new bone may form at right angles to the shaft. Later, a diffusely spreading area of rarefaction develops through the cortex and the medulla as bone destruction becomes apparent.

Pathology

Gross. The tumor extends for a greater distance through the medulla than is apparent in roentgenograms. It extends through and replaces the cortex and often forms a large mass beneath the elevated periosteum. Cut section reveals a grayish-white tumor which is firm and encapsulated by fibrous tissue, sending septae into the tumor and separating it into lobules. Some cystic and necrotic yellowish areas may be visible. Within the subperiosteal tumor, multiple layers of bone lie parallel with the shaft. Some new bone is also deposited around vessels, extending radially from the periosteum to the shaft. The tumor is usually quite vascular and contains hemorrhagic foci. At first, the tumor infiltrates rather than destroys the cortex, usually spreading through the haversian canals. Later, the cortex is destroyed, and the tumor spreads toward the metaphysis and the epiphysis.

Microscopic. The tumor consists of *compactly arranged sheets* of small polyhedral cells with ill-defined borders, meager cytoplasm, and containing nuclei which are large, uniform, prominent, round or oval and possessing scattered chromatin. The nucleus is more than twice as large as that of a lymphocyte. The cytoplasm stains poorly acidophilic. No interstitial stroma can be identified. Many trabeculae of dead bone surrounded by masses of tumor cells almost give the appearance of osteomyelitis. No multinucleated cells are present. The tissue is very vascular. Often, especially in areas of hemorrhage, the tumor cells are clustered about a vessel, and for this reason the tumor has been thought to have an endothelial origin. However, the adventitia of the vessel can be seen interposed between the tumor cells and the endothelium.

Occasionally, a ring of tumor cells form a "pseudorosette," which is similar to the rosette frequently observed in neuroblastoma. However, Ewing's tumor cells tend to accumulate about a center of necrosing tumor cells. On the other hand, the rosette of neuroblastoma is composed of an aggregate of tumor cells about a central core of filamentous neurofibrils.

The tumor cells have a pronounced tendency to degenerate if not fixed and examined soon after removal for biopsy. The nuclei usually shrink and appear pyknotic. This explains the discrepancies between Ewing's original description and that of later observers.

Treatment

The tumor is very sensitive to irradiation, melting quickly, but recurring after several months. Succeeding growths are much less responsive. Therefore, radical surgery plus irradiation is the procedure of choice. The outlook is grave, and any report of a permanent cure must be looked upon with suspicion.

FIBROSARCOMA

Fibrosarcoma of bone is a malignant neo-

plasm, the basic cell of which is the fibroblast in varying degrees of anaplasia. Histologically, it is identical with fibrosarcoma which arises from soft tissue structures, e.g., fascia.

Clinical Picture

Age. Beyond 30

Site of Predilection. Long bones, especially the femur. Also in ribs, skull, vertebrae, mandible

Position. Subperiosteal area of diaphysis or metaphysis; or it originates in medulla and penetrates through to subperiosteal space.

Solitary lesion

Symptoms and Findings. Gradual onset of continuous pain, worse at night. Gradual appearance of swelling, which is smooth, firm, rubbery textured, firmly fixed to underlying bone. Occasionally, tumor is infiltrative and fixes overlying soft tissues, thereby restricting joint motion.

Pathologic fracture occasionally

Metastasizes to the lungs. May metastasize to other subperiosteal spaces.

Roentgenologic Findings

A soft tissue shadow slightly denser than muscle is revealed by soft tissue technic. The shadow is usually a single lesion, extraosseous but immediately adjacent to the cortex of a long bone. Directly beneath the tumor shadow is a saucer-shaped cortical erosion of varying depth. At the upper and the lower corners of the periosteal shadow, slight triangles of reactive bone (Codman's triangles) may be found, particularly in slower growing tumors. Slight calcific densities are sometimes seen within the shadow The tumor is generally large in contrast with the amount of bone destruction. If the tumor is very malignant and infiltrative, the sharp borders of the shadow are lost.

When the tumor has penetrated the cortex, the latter is riddled with multiple small lytic areas The medulla is then involved and shows vague areas of rarefaction

When the tumor rarely originates within the medulla, a central irregular moth-eaten area of rarefaction appears. However, no reactive bone density is seen about it, and the cortex, although rarefied locally, is not expanded.



Fig. 201. Fibrosarcoma.

Eventually, it forms a shadow external to the bone.

Pathology

Gross. The tumor is a well-encapsulated, firm, white, fibrous, glistening mass beneath the elevated periosteum on one side of a long bone. On cut section, strands of fibrous tissue are arranged in striations, whorls, or criss-crossed. The cortex may be uninvolved, and the tumor may peel away easily from the bone. If the tumor has invaded the cortex, the tumor is separated with difficulty, and the underlying bone presents a saucer-shaped depression of varying depth. When the tumor is markedly invasive, the cortex is thoroughly destroyed, the medulla infiltrated, and the bone markedly weakened so that a pathologic fracture occurs. Usually, the tumor grows by expansion and penetrates the periosteum at a late stage to enter the surrounding soft tissues. A very malignant tumor is more infiltrative, destroys and easily spreads through the periosteum, engulfing the overlying structures. The main mass of tumor may occupy the interior of the bone, destroying and replacing both cortex and medulla.

Microscopic. The *most malignant* type of

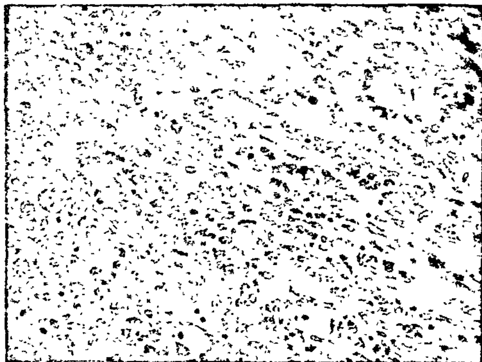


PLATE 29. Fibrosarcoma. The cells are grossly irregular in size and shape, are mainly composed of *spindle* cells and contain occasional dark-staining nuclei. Mitoses are frequent, and intercellular material is sparse. The presence of tumor giant cells differentiates this from a nerve sheath sarcoma. (× 360)

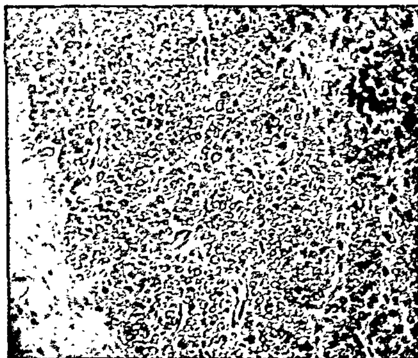


PLATE 30. Reticulum cell sarcoma. The cellular appearance strongly resembles that of Ewing's sarcoma, but intercellular stroma is very much in evidence and reticulin fibers can be demonstrated by specific stains. (× 220)

fibrosarcoma consists mainly of a very cellular tissue composed of small *oat-shaped cells* resembling mesenchymal cells. These cells have scanty cytoplasm and contain small, dark, rounded nuclei with mitoses very much in evidence. The cells are packed tightly so that very little intercellular substance is seen. They are considered to be the least differentiated form of fibroblasts. A *less malignant* fibrosarcoma is one in which the cells are more differentiated. The preponderant cell is the *spindle cell*. This is larger, contains more cytoplasm, and the nucleus is ovoid and vesicular in appearance. These cells, too, are packed tightly, but more intercellular tissue fibers are apparent. Cells and fibers are arranged in fasciculi or bundles, typically in *whorls*, but may also form *palisades* and *criss-crossings*. The *least malignant* tumor is composed chiefly of *fibroblasts and more intercellular material* of the eosinophilic collagenous type. It resembles a nerve sheath sarcoma, but *tumor giant*

cells are present. Each tumor frequently contains all 3 types of cells. The degree of malignity is suggested by the preponderant cells, but this is not absolutely true. No tumor new bone is formed.[42]

Origin of Fibrosarcoma

Most authorities agree that fibrosarcoma originates from the fibrous layer of the periosteum, in a fashion identical with fibrosarcoma starting in other fibrous tissues. Because fibrous tissue occurs in the medulla, the tumor is thought by some observers to start also within the bone.[43, 44, 45] Others believe that fibrosarcoma within the medulla is in reality an osteogenic sarcoma in which ultimate osteogenesis has not occurred. However, true fibrosarcoma never forms tumor bone at its original site or in its metastases.

Treatment

Theoretically, one can differentiate between a benign and a malignant growth. However, a fibroblastic innocent-appearing tumor can be very destructive and metastasize early. Refinements of microscopic diagnosis should not enter into the decision. Amputation offers the only hope of cure. In general, the fibrosarcomas are less malignant than the osteogenic sarcomas.

RETICULUM CELL SARCOMA
(Atypical Ewing's Sarcoma)

All blood-forming tissues have a framework of reticular fibers and cells. These fibers are sheathed by a thin layer of protoplasm in which are scattered pale oval nuclei. These are the *primitive reticular cells* which show no cell limits. They are not actively phagocytic. However, they are capable under certain conditions, as toxic or inflammatory, of transforming into all types of blood and connective tissue cells They transform into large active phagocytic reticular cells, the *fixed macrophages,* which have an abundant cytoplasm, a large pale nucleus, are stellate or spindle-shaped and adhere to the reticular fibers. They contain the debris of dead cells and foreign materials and are capable of ingesting certain dyes as lithium carmine. They may become *free macrophages*. Macrophages are often flattened and resemble endothelial cells. However, true endothelial cells are incapable of ingesting dyes. This dye method may be used to distinguish macrophages from true endothelial tissue. Reticular fibers are best displayed by the silver impregnation method of Hortega.



Fig. 202. Reticulum cell sarcoma. Appearance of tissue stained specifically for reticulin fibers.

The reticuloendothelial cell thus has a pluropotentiality which explains the formation of several types of tumors, notably Ewing's tumor and reticulum cell sarcoma.

Reticulum cell sarcoma is a tumor similar in clinical course, pathologic appearance and radiosensitivity to Ewing's tumor but having a more favorable prognosis.[46-49]

[42] Ewing, J A review of the classification of bone tumors, Surg, Gynec & Obst. 68 971, 1939

[43] Budd, J W, and MacDonald, I Osteogenic sarcoma a modified nomenclature and review of 118 five-year cures, Surg, Gynec & Obst 77 413, 1943.

[44] Coley, B L Neoplasms of Bone and Related Conditions, New York, Hoeber, 1949

[45] Phemister, D B · Cancer of bone and joint, J A M A 136 545, 1948.

[46] Parker, F, Jr, and Jackson, H, Jr.. Primary reticulum cell sarcoma of bone, Surg, Gynec. & Obst 68.45, 1939

[47] Sherman, R S, and Snyder, R E The roentgen appearance of primary reticulum cell sarcoma of bone, Am. J. Roentgenol. 58·291, 1947.

[48] Valls, J, Muscolo, D, and Schajowicz, F.: Reticulum-cell sarcoma of bone, J Bone & Joint Surg 34B 588, 1952.

[49] Khanolkar, V. R Reticulum cell sarcoma of bone, Arch Path. 46 467, 1948.

Clinical Picture

Age. All ages, usually between 20 and 50

Site of Predilection. Long bones chiefly

Position. In medulla or epiphysis or metaphysis

Symptoms. Typically, do not alter the general condition of the patient. Pain usually initial symptom, mild to moderate, never severe Soft swelling. No fever or leukocytosis. Metastases rare, usually to lungs, rarely to other bones

Course. Onset insidious, progression very slow Metastases late.

Pathologic fracture common

Roentgenologic Findings

The tumor is osteolytic. The area of *decreased density* appears in the medulla of the metaphysis or the epiphysis and spreads rapidly to the cortex, then invades the soft tissues. There is notable *absence of reactive bone formation* either in the cortex or the periosteum. Occasionally, residual bone trabeculae within the area of destruction may give a loculated appearance to the tumor. A large area of the bone is rapidly affected. Surprisingly, symptoms are minimal at this time and the general condition of the patient is good.

Pathology

Gross. The tumor is grayish, and its consistency varies between firm and friable. It is vascular and exhibits small areas of hemorrhage. The periosteum is elevated without reactive bone formation. The cortex is thinned, distended and perforated at several points. The tumor tissue extends over a large area in the medulla, the cortex and the subperiosteal space.

Microscopic. The tissue is practically identical with reticulum cell sarcoma of lymph nodes. There is an intense proliferation of the cellular elements. The cells possess slightly basophilic cytoplasm with poorly defined borders, sometimes appearing amoeboid, but definitely ramified, throughout the tissue, by cytoplasmic processes. The nuclei are large, oval, lobulated, or reniform and are poor in chromatin. Blood vessels are intimately associated with the tumor cells, which occasionally are seen invading the vessel wall. By staining with Hortega's technic, a characteristic reticular fibrillar network is displayed. Starting at the adventitia of blood vessels as a close mesh, it gradually becomes more open as it spreads throughout the tissue. However, it maintains a constant effect of enclosing small groups of tumor cells simulating areolae.

In contrast, Ewing's tumor exhibits a compact mass of cells of uniform shape, with rounded or oval nuclei and ill-defined and scanty cytoplasm. The reticulum fibers, when exposed by silver impregnation, surround much larger areas of cells, forming lobules within which the fibrillae do not penetrate.

Treatment

Primary reticulosarcoma is very radiosensitive, but recurrence is relatively frequent. Therefore, a wide excision of the growth, even amputation, plus irradiation will produce a high percentage of cures.

DIFFERENTIAL DIAGNOSIS

	Primary Reticulum Cell Sarcoma	Ewing's Sarcoma
Age	20 to 50	5 to 16
Site	Epiphysis or metaphysis	Metaphysis and diaphysis
Roentgenograms	Osteolytic, no reactive bone, no tumor bone	Initial bone condensation Later osteolytic Onion-peel periosteal bone
General Condition	Usually little altered	Seriously affected
Fever	Absent	Frequent
Metastases	Rare and distant	Frequent and early
Course	Slow	Rapid
Prognosis	High number of cures	Fatal in nearly all

MULTIPLE MYELOMA

Multiple myeloma is a neoplasm consisting basically of cells resembling plasma cells plus cells similar to immature myelocytes; it characteristically invades and replaces cancellous bone slowly in older individuals.

CLINICAL PICTURE

Age. 40 to 60, especially males

Symptoms. Early silent. Gradually pain, at first mild, intermittent and vague, frequently in lumbar and sacral areas, chest and ribs. A severe attack of sharp pain may be brought on by a sudden movement or exertion, as a lifting strain. It may be neuritic, girdlelike at level of chest or referred to a lower extremity. The pain gradually lessens and may disappear. Attacks may recur with increasing frequency. Gradual loss of weight and general debility

Findings. Early, none. Gradually, a diffuse tender swelling of the bone is palpable, particularly noted over sternum and ribs, where multiple nodules may exist. Vertebral collapse results in flattening of lumbar and increased rounding of thoracic spine. Muscle spasm and rigidity over involved vertebra. Pallor of secondary anemia.

Course. Insidious onset, rheumatoid pains usually in back and loins, intermittent and progressive over 3 years, then persistent over subsequent years, eventually rapid in final stages and fatal.

Complications. Pathologic fractures, particularly of the ribs, are frequent; union delayed. Spinal cord or nerve root compression.[50] Secondary anemia. Leukopenia in late stage.

Laboratory Findings. *Bence-Jones proteins in urine.* This is specific for any pathologic process which extensively involves the marrow. On boiling the urine, a white precipitate appears at about 50° C., then dissolves at the boiling point, especially after acidifying the urine. On cooling, the precipitate reappears. If serum albumin is present, the albumin is filtered off at the boiling point, and the procedure is repeated. It is a constant finding only in the late stages. The *serum globulin is increased,* and the *albumin-globulin ratio is reversed. Secondary anemia.* Presence of *immature myelocytic forms* in the blood stream. *Leukopenia* in the late stage after *early leukocytosis.*

ROENTGENOLOGIC FINDINGS

Early the roentgenogram findings are nega-

[50] Davison, C., and Balser, B. H.: Myeloma and its neural complications, Arch. Surg. 35:913, 1937.



FIG. 203. Multiple myeloma, showing multiple round osteolytic defects without alteration in contour of the bone.

Clinical Picture

Age. All ages, usually between 20 and 50

Site of Predilection. Long bones chiefly

Position. In medulla or epiphysis or metaphysis

Symptoms. Typically, do not alter the general condition of the patient. Pain usually initial symptom, mild to moderate, never severe. Soft swelling. No fever or leukocytosis. Metastases rare, usually to lungs, rarely to other bones

Course. Onset insidious, progression very slow. Metastases late.

Pathologic fracture common

Roentgenologic Findings

The tumor is osteolytic. The area of *decreased density* appears in the medulla of the metaphysis or the epiphysis and spreads rapidly to the cortex, then invades the soft tissues. There is notable *absence of reactive bone formation* either in the cortex or the periosteum. Occasionally, residual bone trabeculae within the area of destruction may give a loculated appearance to the tumor. A large area of the bone is rapidly affected. Surprisingly, symptoms are minimal at this time and the general condition of the patient is good.

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Prognosis	High number of cures	Fatal in nearly all

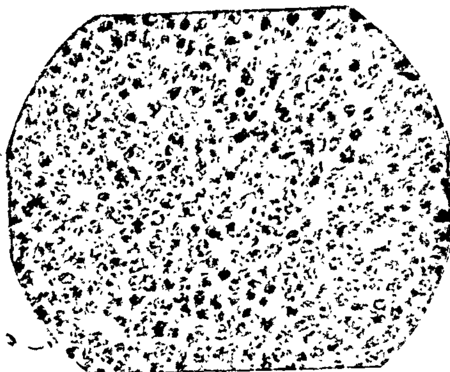


PLATE 31. Multiple myeloma (× 490, oil immersion). Exhibiting closely packed typical myeloma cells with rounded eccentrically placed nuclei and spoke-wheel arrangement of chromatin.

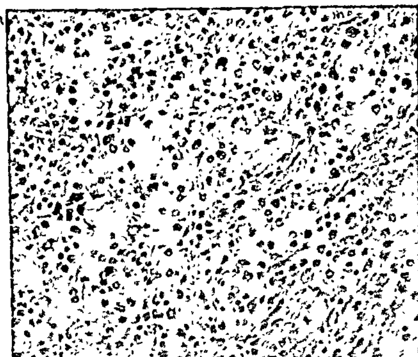


PLATE 32. Liposarcoma (× 415). This section shows a profusion of lipoblasts. Under higher magnification, features to be sought include signet-ring cells, giant-sized cells containing lipoid vacuoles and abundant pink granules, and areas of malignant fibroblasts.

and shapes, abundant mitoses and hyperchromatism. No fat droplets are contained in these cells which make up the bulk of the tumor.

The irregular islands of mucoidlike tissue contain fat cells. These consist of *large adult fat cells* with the usual peripheral position of the nucleus; *smaller young fat cells* with nuclei, either central or peripheral, small, rounded and dark, or very large and occupying most of the cell; fusiform cells with hyperchromatic ovoid nucleus, and cytoplasm vaculated like fat cells. This last type of cell seems to become more atypical, loses its fat content and assumes the appeaarnce of the main tumor cells.

The existence of a liposarcoma primary in bone is controversial, but the histologic appearance and the distinctive clinical course seem to point it out as a distinct entity.

tive. This is especially true with extensive involvement of the spine. The cortex eventually is resorbed in localized fashion and circumscribed, punched-out rarefactions are visible. The cortex *is not expanded, and no reactive* bone is seen, either about the lytic lesions or in the subperiosteal space. These findings are best seen in the skull.

PATHOLOGY[51-54]

Gross. The tumor tissue is a grayish red, or dark-red, soft gelatinous mass which bleeds freely. It lies chiefly within the medullary portion of the bone and where the cortex has been eaten away. The overlying cortex is thin and easily broken No bone trabeculae exist within the tumor tissue The neoplasm erodes and extends beyond the cortex. In the spinal canal it compresses the spinal cord and the *nerve roots.*

Microscopic. The typical cells are round and contain an eccentrically placed nucleus with a nucleolus The chromatin is sparse and arranged in spokes-of-wheel fashion. The nuclear membrane is *well defined.* The *cytoplasm* is eosinophilic. However, the perinuclear halo typical of plasma cells is not seen; and the plasma cell stain, polychrome methylene blue, is not well taken. The tumor cell may represent lack of complete differentiation. Other cells resembling lymphocytes and myelocytes may represent stages in differentiation from the stem cell to the plasma cell. The cells are closely packed, and no supporting stroma is visible. Thin-walled blood vessels pervade the tumor. Fat cells, *megalokaryocytes and eosinophils* are present. The tumor cells surround and directly eat away spicules of bone.

Associated Pathology. Pathognomonic *changes in the kidneys* consist of plugs of protein within the tubules, surrounded by foreign body giant cells. This leads to tubular atrophy, interstitial scarring and renal insufficiency. Generalized bone resorption results in metastatic *calcium deposits* in kidneys, lungs and other tissues. *Amyloidosis* occurs in parenchymatous organs and within the neoplastic tissue. *Neurologic* manifestations ensue from compression of the cord and the nerve roots. Nodules in the lungs produce *pulmonary* complications, as bronchitis and emphysema.

TREATMENT

The widespread dissemination and the eventual fatal outcome make treatment essentially palliative. The tumor is radiosensitive. This relieves pain, but the tumor eventually loses its susceptibility. Temporary relief of bone pain may be obtained by giving Stilbamidine and Pentamidine in conjunction with a high protein diet.[55] Radioactive isotopes, such as P^{32} and Sr^{89}, may be tried, but their value is uncertain.[56]

LIPOSARCOMA

Liposarcoma is a rare nepoplasm of bone resembling a fibrosarcoma in appearance but displaying histologic evidence of origin from fat cells. It is characterized clinically by osteolytic destruction, predilection for the extremities of long bones, destruction of the cortex with spread into contiguous soft parts, a slow course with eventual spread to other bones, and finally visceral metastases. It is radiosensitive.

PATHOLOGY

Gross.[57] The tumor is soft, grayish-yellow, lobulated and coarsely fascicular, resembling fibrosarcoma. It replaces bony substance, perforates the cortex, and a lobulated tumor mushrooms out into the surrounding tissues It displays islands of glistening, opaque mucoidlike tissue.

Microscopic. Interlacing spindle cells resemble those of medullary fibrosarcoma but are more blunt and the cytoplasm more acidophilic. Its malignant character is revealed in central nuclei, which display bizarre sizes

[51] Geschickter, C. F, and Copeland, M M Multiple myeloma, Arch. Surg. 16 807, 1928

[52] Kolodny, A.: Bone sarcoma, Surg, Gynec. & Obst. 44:126, 1927.

[53] Lichtenstein, L, and Jaffe, H. Multiple myeloma, Arch. Path 44 207, 1947

[54] Rosenthal, N., and Vogel, P Value of sternal puncture in multiple myeloma, J Mt Sinai Hosp. 4:1001, 1938.

[55] Snapper, I.: Stilbamidine and Pentamidine in multiple myeloma, J A M A. 133.157, 1947.

[56] Lawrence, J. H, and Wasserman, L. R.: Multiple myeloma treated with radioactive isotopes, Ann. Int. Med 33 41, 1950

[57] Stewart, F. W.: Primary liposarcoma of bone. Am. J Path. 14:621, 1938.

teristic intracellular and extracellular vacuoles of mucin. The cells resemble bladder epithelium and are arranged in cords, columns or clusters.

Clinical Picture

The following relates only to sacrococcygeal chordoma.

Symptoms are due to expansion and bone destruction. At first, pain occurs in the rectal and anal regions and is mild and intermittent. Obstinate constipation and urinary difficulty develop. Later, incontinence is followed by motor and sensory disturbances of the lower extremities, the gluteal region and the external genitalia. Finally, pain becomes intractable, the lower extremities display muscle weakness, and fecal and urinary incontinence is severe. The course is slow but progressive.

Examination reveals a tender, soft, smooth, fixed mass over the posterior and/or anterior aspect of the sacrum.

Roentgenologic Findings

A well-circumscribed osteolytic expansile defect is observed in the lower sacrum. A



Fig. 204. Hemangioma of bone. The vessels appear as blood vessel spaces lined by a single layer of flattened endothelial cells. Vessel walls lack elastic fibers and smooth muscle. Connective tissue between vessels is sparse. At the upper edge of this section, bone is being eroded in some areas by vessels, and in others active bone formation is taking place. This is the cavernous type of hemangioma.

ADAMANTINOMA

Adamantinoma of long bones is a rare tumor which is similar to the growth appearing more commonly in the mandible and characterized by epithelial components supposedly derived from enamel-producing cells. The number of cases reported is inadequate for determining its etiologic characteristics. The greatest age incidence seems to be between 10 and 35, and the colored race is predisposed. The tibia is the favored location.[58]

Clinical Picture

The tumor develops insidiously over a period of years, and the initial symptom varies, including pain, swelling, tenderness and pathologic fracture. It is locally invasive, spreading slowly but progessively without metastasizing even after unsuccessful removal.

Roentgenologic Findings

Rarefaction in a medullary location and involving the overlying cortex is the main finding. The lesion is small at first, but with growth the cortex becomes attenuated and expanded, and a honeycombed or loculated appearance is typical. Almost no periosteal reaction is noted. The tumor attains a large size and comes to occupy a major portion of the shaft. The shaft becomes widened and deformed, and eventually the cortex is penetrated with extension into the contiguous soft tissues.

Pathology

Gross. The appearance varies, but most often the tumor is gray or grayish-white and firm in consistency.

Microscopic. Three basic patterns are observed:

1. Masses of epithelial islands in which the peripheral cells are columnar and arranged in palisade fashion. The central cells of these masses are stellate and produce a reticulum appearance.
2. Islands of basal cells with peripheral palisading and central cystic degeneration
3. Islands of squamous epithelium and pearl formation

The stroma is fibrous. Islands of spindle cells may be a feature.

[58] Baker, P. L., Dockerty, M B, and Coventry, M. B.: Adamantinoma (so-called) of the long bones, J. Bone & Joint Surg 36A:704, 1954

Treatment

The tumor is highly radioresistant and frequently recurs after removal. Perhaps one attempt at excision may be justified, but recurrence is an indication for amputation. Years of needless disability may be avoided thereby.

CHORDOMA

A chordoma is a rare neoplasm of the cranium and the spine apparently derived from embryonic remnants of the notochord. The notochord originates from the entoderm and persists as small remnants in the nucleus pulposus of the intervertebral disks. The majority of chordomas arise in the skull about the sphenooccipital region and in the sacrococcygeal region. Their location in other areas of the spine is exceedingly rare. Sphenooccipital tumors favor young adults, and sacrococcygeal tumors most often develop after the age of 40.[59] However, no age is immune.

Pathology[60]

Gross. The tumor is a large, round, smooth, soft gelatinous mass firmly fixed to the bone and locally invaded tissues. The bone is destroyed and replaced by the tumor and, at first, the cortices are thinned and expanded outward. The lower half of the sacrum is usually involved by sacrococcygeal tumors. Growth and extension of the tumor are slow, bulging the sacrum posteriorly and anteriorly before penetrating the cortex and extending into the soft tissues. Within the pelvis, the tumor involves the lumbosacral plexus, the rectum, the bladder and the genital organs. Metastases to the regional lymph nodes occur very late, usually after a period of years.

Cranial chordomas likewise develop slowly, destroy and replace bone and invade intracranial structures.

Vertebral chordomas develop first within the body and extend to the arch, with eventual collapse of the vertebra. Several adjacent vertebrae may be destroyed.[61]

Microscopic. The tissue consists of large, polyhedral cells with small nuclei and charac-

[59] Mabrey, R E: Chordoma a study of 150 cases, Am J Cancer 25 501, 1935

[60] Gentil, F., and Coley, B L: Sacrococcygeal chordoma, Ann Surg 127·432, 1948.

[61] Wood, E H., Jr., and Himadi, G M.: Chordoma. roentgenologic study, Radiology 54:706, 1950.

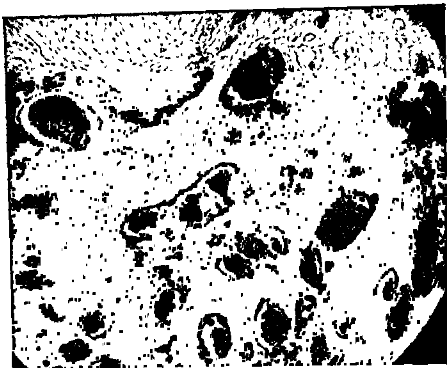


PLATE 35. Hemangioma of bone. The vessels appear as blood spaces lined by a single layer of flattened endothelial cells. Vessel walls lack elastic fibers and smooth muscle. Connective tissue between vessels is sparse. At the upper edge of this section, bone is being eroded by pressure of adjacent vessels, whereas in other areas active bone formation is taking place. (× 70)

be attempted, but recurrence almost inevitably occurs.

HEMANGIOMA OF BONE
(Angioma of Bone)

Hemangioma of bone is a slowly growing benign tumor composed of masses of fully developed adult blood vessels. They occur frequently as silent lesions in any portion of the skeletal system and other organs. About 12 per cent of spines contain the lesion. However, seldom does it become symptomatic and then only because of pressure effects. A hemangioma has often been thought to cause spontaneous hemorrhage, the resultant hematoma antedating the development of a bone cyst.[63]

ETIOLOGY

Two theories are held:

1. **Congenital Hamartoma.**[64] A vast plexus of primitive vascular channels exists in the embryo. Those not forming part of the circulatory system are obliterated. If they persist, they constitute congenital malformations.

2. **Neoplasm.**[65, 66] Congenital rests by a process of endothelial proliferation differentiate into blood vessels.

PATHOLOGY[66, 67]

Gross. The tumor consists of masses of blood vessels in a fibrous stroma. The adjacent bone is resorbed by expanding pressure, and new reactive bony trabeculae form. The periosteum is not perforated. There is a tendency to extensive thrombosis, organization, regression of the lesion, and healing. When a vertebra is involved, particularly in the normally narrow dorsal area, enlargement of the vertebra or extension into the vertebral canal causes compression myelitis and radiculitis.

Microscopic. Two types are identified—capillary and cavernous. The *capillary type* consists of closely packed small blood vessels lined by a single layer of nearly cuboidal endothelial cells.

The *cavernous type* is more common. It is composed of a multitude of large blood-filled spaces lined by a single layer of flattened endothelial cells. No elastic fibers or smooth muscle can be observed in vessel walls. Connective tissue between vessels is sparse and

[63] Ritchie, G., and Zeier, F. G.: Hemangiomatosis of the skeleton and spleen, J. Bone & Joint Surg 38A:115, 1956.

[64] Schafer, P W.: Pathology in General Surgery, pp 26-31, 120, Chicago, Univ. Chicago Press, 1950.

[65] Ribbert, H · Geschurulstlehre, p 201, Bonn, Friedrich Cohen, 1914

[66] Thomas, A · Vascular tumors of bone, Surg, Gynec & Obst. 74:777, 1942

[67] Bucy, P. C.. Hemangioma of bone, Am. J. Path. 5·381, 1929.



PLATE 33. Adamantinoma. This shows one basic pattern of epithelial islands with peripheral columnar cells in palisade fashion and central reticulum formation. The stroma is fibrous. (× 84)

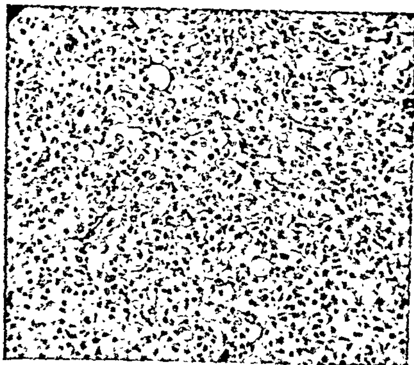


PLATE 34. Chordoma. (× 160)

rounded soft-tissue shadow extends anteriorly and posteriorly. In the case of rare vertebral involvement, one or several adjacent vertebral bodies and their arches are destroyed and collapsed.

Differential Diagnosis

Chordoma of the sacrococcygeal region must be differentiated from chondrosarcoma, tuberculosis, tumors of the female pelvic organs, sacrococcygeal teratoma and rectal carcinoma.

Treatment

Although generally regarded as radiosensitive, the tumor has been eradicated successfully by special rotation technics employing extremely high voltages.[62] Chordomas in children are more radiosensitive than in adults. Otherwise, complete surgical excision should

[62] Friedman, M., Hine, G J, and Dresner, J: Principles of supervoltage rotation therapy, illustrated by treatment of a chordoma of a vertebra, Radiology 64 1, 1955.

composed of a *mass of anastomosing vascular channels*. The vascular walls are formed by the tumor cells, which are *large polyhedral cells* with a well-defined cell membrane and *pale vesicular nuclei* with dusty chromatin. The *cells are several layers thick* in contrast with the benign angiomatous tumors in which a single cell thickness is found. When many vessels are seen in cross section, an alveolar appearance results and may be confused with metastatic hypernephroma. However, the latter's cells are large, cylindrical, have small nuclei, and the lumens are empty. Hemangioendothelioma tumor cells often freely invade blood vessels and flourish as intravascular tumor thrombi.

Occasionally, the tumor is densely cellular and almost fibrosarcomatous in appearance. However, many small blood vessel spaces lined by and surrounded by multiple layers of endothelial cells can be identified.

The histologic picture varies even in the same case from time to time, depending on the rapidity of cellular growth and differentiation into vasoformative elements. The diagnostic feature is always the formation of new blood vessels. Typical endothelial cells should be identified.

Treatment

The tumor is only slightly radiosensitive. After radiation therapy, a malignant tumor displaying hypercellularity, pleomorphism, mitoses and disorderly appearance may revert to a benign appearance resembling a capillary hemangioma. The change is temporary, and malignant characteristics reappear. Radical surgical excision by amputation is recommended, but the outlook is poor.

IDENTIFICATION OF ENDOTHELIAL TISSUES

The literature is replete with the term "endothelioma," which has caused confusion in classifying various tumors. A tumor labeled as an "endothelioma" should contain a considerable number of cells which can positively be identified as endothelial cells. These cells possess certain histologic characteristics,[76] and definite evidence of their vasoformative potential should be present.

During embryonic development, the squamous endothelial cells arise through flattening of mesenchymal cells. Mesenchymal cells are smaller than fibroblasts but have the same general appearance. They are outstretched, stellate cells with a large, oval, pale, vesicular nucleus containing dustlike chromatin particles. The endothelium of blood vessels may have identical potencies with mesenchymal cells, i.e., they are capable of differentiating into various blood and macrophagic elements. Also, the endothelium has the potential of reverting to mesenchymal cells, and vice versa.

When connective tissue fibrils appear between mesenchymal cells, the tissue becomes connective tissue. The appearance of fibroblasts is almost identical in appearance with mesenchymal cells. Therefore, undifferentiated mesenchymal cells may be considered to be present at all times within connective tissue. These are observed particularly about capillaries.

Endothelial cells are structurally similar to fibroblasts and mesenchymal cells. The elongate oval nucleus is flattened and contains fine dustlike chromatin particles. *However, it lacks the large nucleoli.* Its membranes often show longitudinal folds. The flat endothelial cells are usually stretched along the axis of the capillary and have tapering ends. In wider capillaries they are shorter and broader. In a silver nitrate stain, sharply stained black boundaries of cells are demonstrable.

Great numbers of capillaries are collapsed and not visible when the organ is at rest. They open up and are made visible when blood flows through them.

Benign vascular tumors may constitute distention of pre-existing channels or an increased vasoformative potential of undifferentiated mesenchymal cells. Malignant vascular tumors may represent a wild, disorderly, rapid exhibition of vasopotential of mesenchymal tissue. Growth may be so rapid that the tissue is very cellular and does not have time to differentiate into clearly vascular structures; or typical endothelial cells are formed in profusion and tend to arrange themselves in tubules or alveoli whose lumens contain blood. *The one feature which identifies an endothelioma is a recognizable vasoformative tendency.*[77]

[76] Maximow, A. A., and Bloom, W.: *Textbook of Histology*, Philadelphia, Saunders, 1958.

[77] Thomas, A.: Vascular tumors of bone, Surg., Gynec. & Obst. 74:777, 1942.

may be compressed thin. Adjacent bony trabeculae are resorbed, possibly because of vascular tension. In other areas, new reactive bone formation is active.

Clinical Picture

Most lesions are silent and are discovered accidentally. Symptoms are produced by pressure on surrounding structures and may develop at any age. The site of predilection is a vertebra, often between the 12th thoracic and the 4th lumbar. The next favored site is the skull, where the lesion usually originates in the meninges and the scalp. Infrequently, the long bones are involved. Pain constitutes the main local symptom. Hemangioma of a vertebra, especially when midthoracic in location, will produce neurologic manifestations, depending upon the site and the degree of narrowing of the spinal canal. The tumor increases in size very slowly, then may persist or undergo spontaneous regression and healing.

Roentgenologic Findings

These depend upon the type of bone involved:

1. **Flat Bone.** Trabeculae of new bone radiate outward from a central focal point producing a characteristic "sunburst" appearance (plexiform angioma).
2. **Vertebra.** Coarse, dense, vertical striations in the body. These may extend into the vertebral arch. The body becomes ballooned, with rounding of the concave borders
3. **Tubular Bone.** The lesion is usually metaphyseal and eccentric. Multilocular cystic cavities of various sizes and irregular shapes resemble the soap-bubble appearance of a giant cell tumor, but the loculi are smaller. A fine fibrillary network is contained within each locule. A local or general uniform spindle-shaped expansion of the shaft takes place. The cortex is thinned but intact.

Treatment

Surgery is fraught with the danger of uncontrollable hemorrhage. These tumors may regress and heal by moderate dosages of x-rays given at regular intervals over a number of months.[68] However, radiation therapy may damage the spinal cord. A vertebral hemangioma should be destroyed by resection or cauterization.[69]

HEMANGIO-ENDOTHELIOMA[70-75]

(Angio-endothelioma; Hemangiosarcoma)

In this very rare tumor the basic cell is endotheliomatous in appearance; therefore, it might be classified as a variant of a Ewing's tumor. It represents an extremely malignant tumor which often has extended by hematogenous spread before it is recognized.

Clinical Picture

Age. Adults exclusively

Area of Predilection. Reported in long bones and ilium. More common near surface of the body as soft bluish-red tumors with satellite nodules

Symptoms. Pain varies from ache to sharp shooting.

Findings. Deep-seated swelling, soft, tender, pulsating. If it extends to surface, bluish color is apparent. Bruit on auscultation.

Pathologic fracture

Roentgenologic Findings

Single or multiple osteolytic areas without reactive bone. Moth-eaten appearance.

Pathology

Gross. The tumor may appear benign and similar to an ordinary bluish-red hemangioma of bone. The interior is composed of a mass of bleeding vessels.

Microscopic. This bulky, cystic tumor is

68 Meyerding, H W.: Hemangioma of bone, J. Bone & Joint Surg. 18.617, 1936.

69 Bucy, P C., and Capp, C S · Primary hemangioma of bone, Am. J Roentgenol 22 1, 1930

70 Allen, E. V, Barker, N. W., and Hines, E. A: Peripheral Vascular Diseases, Philadelphia, Saunders, 1946

71 Copeland, M. M, and Geschickter, C. F.: Tumors of Bone, Philadelphia, Lippincott, 1949.

72 Gordon-Taylor, G, and Wiles, P.: Pulsating angio-endothelioma of the innominate bone treated by hindquarter amputation, J Bone & Joint Surg 31B·410, 1949

73 Luck, J. V.· Bone and Joint Diseases, Springfield, Thomas, 1950

74 Resink, J. E J: Case of skeletal hemangioendothelioma, Fortschr. Geb. Röntgenstrahlen 80:732, 1954.

75 Thomas, A: Vascular tumors of bone, Surg, Gynec. & Obst. 74:777, 1942.

system[3, 4, 5] and is unrelated to the location of the primary growth. Therefore, it is generally accepted today that true embolic bone metastases are blood borne; furthermore, that "the distribution of metastases in the bones is almost exclusively dependent upon the arrangement of the blood vessels into which tumor emboli are released and by means of which they gain entrance to the capillary beds."[6]

DIRECT NONMETASTATIC INVASION FROM CONTIGUOUS GROWTHS

Meningiomas and craniopharyngiomas, although considered essentially as benign tumors, may erode and invade the bones of the skull. The facial bones, the mandible, the hyoid bone and occasionally the base of the skull and the cervical vertebrae may be invaded by ulcerating cutaneous, oral or pharyngeal carcinomas or by their cervical lymph-node metastases. Carcinoma of the nasal mucosa, especially antrum, may result in extensive destruction and perforation of the adjacent bones. More unusual cases include carcinoma of the ethmoid invading the cranial bones and melanoma of the eye invading the orbital bones. The thoracic skeleton may suffer direct invasion from mammary, pulmonary or esophageal carcinomas. Mammary carcinoma mainly spreads into the adjacent ribs, while carcinoma of the esophagus often invades the vertebral bodies. In these cases, ulceration and secondary infection of the cancerous growths markedly accelerate bone destruction (Fig. 205). In noninfected tumors, the periosteum or perichondrium constitute a most effective barrier to tumor extension. Pelvic bones may be invaded by carcinomas of the urinary bladder or of the uterine cervix. Rarely, an ovarian carcinoma or a carcinoma of the rectum may invade the sacrum and the coccyx by contiguity. Other tumors reported in the literature as invading the bones are carcinoma of the salivary glands invading the mandible; chordoma invading the sacrum, the vertebrae, or the base of the skull; sarcoma of adjacent soft tissues, chondrosarcoma and synovioma invading the long bones.



Fig. 205. Metastatic carcinoma of the lumbar vertebrae with secondary infection (primary in the urinary bladder). Note extension into the intervertebral disk.

Direct bone invasion from contiguous growths has little clinical importance. As a rule, these tumors do not present difficult diagnostic problems, and their spread is usually beyond the reach of therapeutic measures. The symptoms are dependent upon the location of the primary growth, its extension and the structures involved.

BONE METASTASES

Blood-borne metastases to the bones pre-

[3] Batson, O V · The role of the vertebral veins in metastatic processes, Ann Int. Med. 16:38-45, 1942.

[4] Coman, D R, and de Long, R. P · The role of the vertebral venous system in the metastasis of cancer to the spinal column, Cancer 4 610-618, 1951.

[5] Prinzmetal, M, Ornitz, E M, *Simkin, B*, and Bergman, H. C · Arteriovenous anastomoses in liver, spleen and lungs, Am J. Physiol 152:48-52, 1948

[6] Coman, D, R, de Long, R P, and McCutcheon, M.· *Studies* on the mechanisms of metastasis; the distribution of tumors in various organs in relation to the distribution of arterial emboli, Cancer Res. 11 648-651, 1951.

14

Secondary Tumors of Bones

RENATO BASERGA, M.D.*

Secondary neoplastic growths of bones, as the name implies, are cancerous tumors, originating in other organs and involving the skeletal structures of the body.

Bones may become involved by secondary tumors in two ways: (1) by direct invasion from contiguous growths, and (2) by blood-borne metastases. According to Willis' definition, metastasis is a secondary growth originating from a detached tumor fragment.[1] Thus, invasion by a contiguous growth cannot be considered a true metastasis. It should also be mentioned that some authors have suggested the possibility of bone metastases by the lymphatic channels.[2] However, the following circumstances are against the possibility of lymph-borne metastases:[1] (1) Lymphatics have not been demonstrated in bone marrow. (2) Cancerous permeation of the lymphatics of the periosteum is found rarely, and solely at the level of the foramina. (3) Metastases in bones and in adjacent lymph nodes coexist in only a minority of cases and such coexistence is often clearly accidental. (4) There is indubitable evidence that the distribution of metastatic growths in bones depends upon the normal or pathologic distribution of the red bone marrow and the peculiarities of the venous

* Original tables, microphotographs and photographs of gross specimens are from the Museum of Pathology, St Luke's Hospital, Chicago

[1] Willis, R A The Spread of Tumours in the Human Body, St. Louis, Mosby, 1952

[2] Hodges, P C, Phemister, D B, and Brunschwig, A · The roentgen-ray diagnosis of diseases of bone *in* Ross Golden's Diagnostic Roentgenology, New York, Nelson, 1941

TABLE 6. INCIDENCE OF SKELETAL METASTASES ACCORDING TO THE LOCATION OF THE PRIMARY GROWTH*†

SITE OF PRIMARY GROWTH	NUMBER OF CASES	NUMBER OF CASES WITH SKELETAL METASTASES	PERCENTAGE
Mammary gland	85	40	47 0
Prostate	42	19	45.2
Kidney	31	9	29 0
Lung	187	43	22.9
Thyroid	9	2	22.2
Liver	21	4	19 0
Urinary bladder	45	7	15.5
Uterine cervix	34	5	14.7
Pancreas	42	4	9.5
Stomach	108	8	7.4
Large bowel	166	10	6 0
Other sites	225	25	11.1

* Excluding lymphomas

† This material, collected at autopsy at St Luke's Hospital, Chicago, in a 15-year-period (1940-1954) consisted of a total of 1,237 cancerous growths From this group were excluded 42 cases of leukemia and multiple myeloma and 137 cases of brain tumors, which do not metastasize. Among the remaining 1,058 cases, 213 had metastases in the bones for an incidence of 20 1 per cent

dence of skeletal metastases are still valid. These sites are vertebrae, ribs, pelvis, proximal ends of femur and humerus, sternum and skull vault[10] (Table 7). These are the locations in which red marrow is found in the adult.

Ordinarily, metastases are uncommon below the knees or the elbows, but numerous exceptions do occur[11] (Fig. 206). Cases have been reported of metastatic epidermoid carcinoma of the lung in a terminal phalanx, carcinoma of the breast in small bones of the feet, carcinoma of the cervix in the lower end of the tibia, and carcinoma of the colon in the bones of both hands and feet.

OUTLINE OF SYMPTOMS AND DIAGNOSIS

Clinically, pain, pathologic fractures and anemia are important features. The location, the frequency, the radiation and other characteristics of the pain will depend upon the bone involved. When the skull is involved, headache may result. Metastatic growths of the spine, by compressing the nerve roots or the spinal cord, cause girdle pains and neurologic manifestations Numbness of the legs or the arms, objective loss of sensation, spastic paralysis or weakness of the extremities, loss of vesical control and weakness of the rectal sphincter are observed. Metastatic deposits in the bones of the extremities produce pain, swelling and tenderness. Metastases in the ribs and the sternum, on the whole, are frequently silent. Small growths may be asymptomatic or may produce mild and transient pain which attracts only little notice. Such cases are often discovered fortuitously during an x-ray study for other diseases. Conversely, when metastases are voluminous or located in an area which is subject to pressure, the onset may be sudden and characterized by intense pain.

Pathologic fractures are frequent especially, it is stated, when the femur is involved, but in our own experience pathologic fractures of the ribs are equally frequent. As the disease progresses and skeletal involvement becomes more widespread, destruction of the vertebral bodies produces impaction and collapse of the vertebrae. However, osteoplastic metastases progress more slowly than the unossified forms, and pathologic fractures are less frequent. In the terminal stages of the disease, emaciation, anemia and pathologic fractures, often with considerable pain, predominate the picture.

Laboratory Diagnosis

Replacement of the bone marrow by the neoplastic process gives rise to a distinctive hematologic picture. The red blood cells show evidence of a progressive anemia, often of the nonmegaloblastic macrocytic type (myelophthisic anemia). The white corpuscles often display myeloid leukocytosis (20,000 to 30,000 white blood cells per cubic millimeter are not unusual). Nucleated erythrocytes and immature granulocytes are present in the peripheral blood.[12] There are also cases of hypochromic microcytic anemia with leukopenia, especially in osteoplastic metastases. It is in these patients with osteoplastic metastases, who exhibit severe anemia and compensatory erythropoiesis, that metastatic growths can be found in those areas in which the yellow marrow has been replaced by hyperplastic red marrow. In late stages, the anemia is aggravated by a hemolytic component.[13] The platelets are frequently deficient in number: hemorrhagic states are not infrequent,[14, 15] and hemorrhages are often prominent in the metastatic tumor tissue. In cases of anemia due to cancerous disease of the bone marrow, heterotopic myeloid hemopoiesis has been observed occasionally in the spleen and in the liver.

Open biopsy and aspiration biopsy offer a valuable aid to diagnosis and are actually the only procedures that can be depended upon in

[10] Walther, H E: Krebsmetastasen, Basel, Schwabe, 1948.

[11] Geschickter, C. F, and Copeland, M. M.· Tumors of Bone, Philadelphia, Lippincott, 1949.

[12] Ley, A B: Mechanisms of anemia in cancer, M Clin. North America 40.857-870, 1956.

[13] Sohier, W. D, Jr., Juranies, E, and Aub, J. C.: Hemolytic anemia, a host response to malignancy, Cancer Res 17:767-774, 1957.

[14] Lawrence, J S, and Mahoney, E. B · Thrombopenic purpura associated with carcinoma of the stomach with extensive metastases, Am J. Path. 10.383-390, 1934.

[15] Smith, W T, and Whitfield, A G W.: Intravascular microembolic carcinomatosis as a cause of purpura, Brit. J. Cancer 8 97-106, 1954.

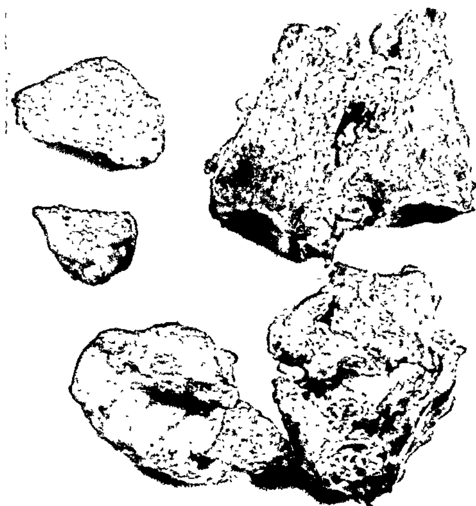


FIG. 206. Metastatic carcinoma of the lower epiphysis of the radius and of the small bones of the hand (primary in the lung).

ponderate numerically over primary bone tumors to such an extent that after the age of 40 every bone tumor must be considered metastatic unless otherwise proved.[7] Still, their incidence is probably greatly underestimated. Table 6 gives the incidence of bone metastases according to the location and the type of primary growth. In other sources, the incidence of bone metastases in malignant disease averaged between 15 and 20 per cent of the cases. The highest incidence (27%) was reported by Abrams and co-workers.[8]

Metastatic growths may appear in any part of the skeleton and even in heterotopic bone. However, von Recklinghausen's conclusions of long ago[9] regarding the sites of maximum inci-

[7] Schinz, H R, Baensch, W. E., Friedl, E., and Uehlinger, E.: Roentgen-Diagnostics (translated by J. T. Case), vol 2, New York, Grune, 1952.

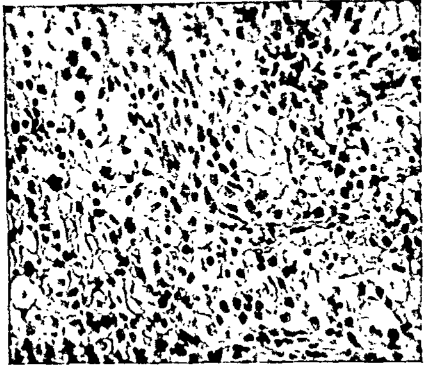
[8] Abrams, H. D, Spiro, R, and Goldstein, N.: Metastases in carcinoma: analysis of 1,000 autopsied cases, Cancer 3:74-85, 1950

[9] von Recklinghausen, F.: Die Fibrose oder deformierende Ostitis, die Oseomalacie und die osteoplastiche Carcinose in ihren gegenseitigen. Beziehungen Festschrift zu Rudolph Virchow's 71 Geburtstage, p. 17, 1903

TABLE 7. DISTRIBUTION AND INCIDENCE OF BONE METASTASES IN 176 CASES OF MALIGNANCY (EXCLUDING LYMPHOMAS)

SITES	NUMBER OF CASES WITH METASTASES
Spine	122
Ribs	88
Skull	21
Pelvis	18
Sternum	18
Femur	13
Humerus	8
Others	12

PLATE 36. Metastatic hypernephroma. (× 360)



growths in the spine are situated almost invariably in the vertebral bodies, rarely in the arches or the processes. Blood-borne periosteal metastases without medullary growths are very rare and then are found only at the level of the foramina. Grossly, bone metastases appear as bone-destroying or bone-producing lesions or a mixture of these two. In the osteoclastic or osteolytic type, metastases appear as well-defined nodules of gray tumor tissue occupying and replacing the cancellous bone and bone marrow (Fig. 207). Cartilage constitutes an effective barrier against tumor invasion. Pathologic fractures are not infrequent, but they may heal by osseous union in a normal fashion even without treatment. In the ribs and in advanced stages, the metastatic tumor may produce a fusiform swelling, involving and replacing an entire segment of bone (Fig. 208). In the osteoplastic type, the affected bone is heavier and denser than normal. Marrow spaces are obliterated by the deposition of new bone, whose structure may be spongy or eburnated (Fig. 209). Thickenings and deformities of the bone due to subperiosteal deposition are present and may result in a diffuse nodular enlargement of the bone. Path-

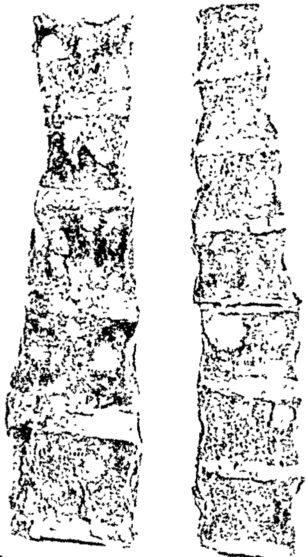


FIG 207. Osteolytic metastases of thoracic and lumbar vertebrae.

determining conclusively the type of growth. However, even open biopsies present often difficult diagnostic problems, especially in locating the primary focus, and some cases must await a thorough postmortem examination for solution [16]

Marrow aspiration has been used in the diagnosis of metastatic bone cancer (Jonsson and Rundles,[17] Mallet[18]). Sites of marrow aspiration are selected by preference in areas of bone pains, tenderness and x-ray abnormality. Marrow sections are much preferable to marrow spreads.[19] Tumor implants are found in about one half of the patients with malignant disease—most frequently in patients with neuroblastoma, carcinoma of the prostate or carcinoma of the breast. Such procedure has been recommended to establish the diagnosis of widespread cancer, especially in cases in which extensive surgery is being contemplated.

Roentgenologic Diagnosis

The great majority of skeletal metastases are not demonstrable roentgenologically.[20, 21] In a minority of cases, they bring about grossly visible bone lesions, of which two types are recognized: the osteolytic, which is more frequent, and the osteoplastic. The latter is not uncommon in metastases from carcinoma of the prostate. Osteolytic lesions appear as irregular or sharply circumscribed areas of reduced density with little evidence of repair. There is practically no periosteal reaction. Mottling with increased density of the bone occurs within the area of destruction, and thickening of the cortex appears above or below the site of metastasis. This reaction is often marked after roentgen therapy over the affected bone. When mottling occurs within an area of destruction in the bone, it favors the presence of metastatic processes as opposed to the more definitely punched-out areas of destruction seen in multiple myeloma.

Osteoplastic metastases appear as areas of increased density, often accompanied by periosteal reaction. The bones assume a mottled or marbled appearance.

A solitary area of metastatic carcinoma must be differentiated from a latent cyst of the bone, a solitary focus of multiple myeloma or the osteolytic form of osteogenic sarcoma. A latent cyst of bone occurs usually in the younger age group, is confined to the interior of the bone, has distinct signs of ossification in the bone shell, and usually symptoms are minimal. A solitary focus of multiple myeloma can be differentiated by the abnormal pattern of the plasma proteins and in some cases by the presence of Bence-Jones protein in the urine. Pulmonary metastases, when present, rule out multiple myeloma. The osteolytic form of osteogenic sarcoma has a greater tendency to be asymmetrically located in the bone, and there is evidence of more rapid destruction and periosteal reaction. If the single bone metastasis is from a renal carcinoma, which is often the case, an examination of the patient will disclose hematuria and other evidence of kidney involvement. Diffuse osteoplastic metastases must be differentiated from Paget's disease. In the latter the serum alkaline phosphatase is markedly elevated, but the serum acid phosphatase is within normal limits. Osteoplastic metastases, either from mammary or prostatic tumors, are accompanied by an elevated acid phosphatase.

OUTLINE OF PATHOLOGY

In most instances of bone metastases the primary tumor is known, but even if we restrict bone lesions of metastatic nature to those in which the diagnosis is obscure, metastatic tumors are still the commonest of all malignant bone neoplasms.[22] Metastatic growths in bones being blood-borne, they are situated initially in the bone marrow.[23] Metastatic

[16] Hirsch, E F.: Pathology in Surgery, Baltimore, Williams & Wilkins, 1953.

[17] Jonsson, U, and Rundles, R W Tumor metastases in bone marrow, Blood 6 16-25, 1951

[18] Mallett, L Hibernation de la cellule cancereuse, Acta Intern Union against Cancer 6 993-1005, 1950

[19] Agress, H Comparative study of spreads and sections of bone marrow, Am J Clin Path 27 282-299, 1957.

[20] Schinz, H. R, Baensch, W E, Friedl, E, and Uehlinger, E: Roentgen-Diagnostics (translated by J. T Case), vol 2, New York, Grune, 1952

[21] Walther, H. E. Krebsmetastasen, Basel, Schwabe, 1948

[22] Ackerman, L. V. Surgical Pathology, St. Louis, Mosby, 1953

[23] Sharpe, W. S, and McDonald, J R.: Reaction of bone to metastases from carcinoma of the breast and the prostate, Arch. Path. 33:312-325, 1942.

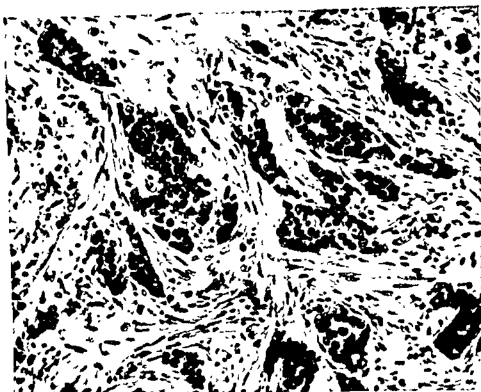


Fig. 210. Proliferation of connective tissue in metastatic carcinoma of bone (primary in the mammary gland). (× 390)

slightly less than the number of investigators. According to Willis,[25] the proliferation of bone accompanying metastatic lesions is similar to the proliferation of the connective tissue stroma in desmoplastic growths or to the glial reaction to tumors invading the central nervous system. Osteoplasia, desmoplasia and glial proliferation are similar expressions of different stromal reactions to neoplastic invasions. In fact, many osteoplastic carcinomas are also desmoplastic. Osteoplastic mammary carcinomas always, and osteoplastic carcinomas of the prostate are often slow, infiltrative growths showing scirrhous characters in soft tissues.

THE PRIMARY TUMORS

Carcinoma of the Mammary Gland

Carcinoma of the breast leads the list with a frequency of bone metastases of about one half of the fatal cases. The bones most frequently involved are spine, pelvis, femur (upper third), skull, ribs and humerus (at the junction of the upper and the middle thirds). The interval between radical mastectomy and actual roentgenographic demonstration of bone metastases is between 15 and 30 months, with an average of 19.8 months. Bone metastases are often preceded by metastases to the regional lymph nodes, and in one third of cases they are accompanied by pulmonary metastases.[26] Clinically, the appearance of metastases is characterized by severe pain which will vary according to the location of the metastases. Pathologic fractures occur in almost half of the cases, more often in the vertebrae, the ribs or the femur. Roentgenologically, from 80 to 90 per cent of the metastatic deposits are of the osteolytic type.[26] The diagnosis is confirmed by laboratory tests. The copper-resistant serum acid phosphatase is not only of diagnostic importance[27] but it is also an indication of the effectiveness of treatment.[28] Regressions or remissions of bone lesions are accompanied by a decrease of the serum level of the acid phosphatase and by a decreased calciuria.

[25] Willis, R A: The Spread of Tumours in the Human Body, St. Louis, Mosby, 1952.

[26] Staley, C. J.: Skeletal metastases in cancer of the breast, Surg., Gynec. & Obst 102:683-688, 1956.

[27] Reynolds, M. D, Lemon, H. M, and Byrnes, W. W.: Copper-resistant serum acid phosphatase. I. Method and values in health and disease Cancer Res. 16:943-950, 1956.

[28] Lemon, H. M., and Reynolds, M. D.: Regression of mammary cancer metastases following decline of serum acid phosphatase induced by anti-estrogen therapy, Proc. Am. A. Cancer Res. 2:129, 1956.



Fig. 208. Metastatic carcinoma of the rib (surgical specimen).

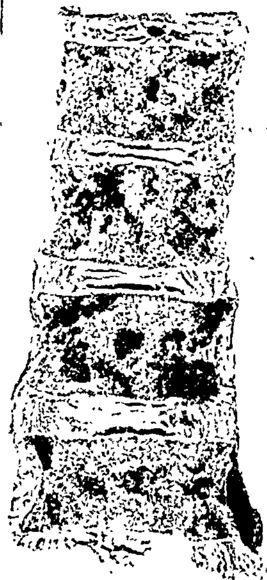


Fig. 209. Osteoplastic metastases of the lumbar vertebrae (primary in the prostate). The dark areas are the residues of marrow spaces.

ologic fractures rarely occur. It should be noted that carcinoma metastases, particularly from the prostate, may be of the osteoplastic type in early stages of the disease, while those developing in the same individual later, and especially in the terminal stages, may be entirely of the osteolytic type.

Histologically, it should be emphasized that no sharp distinction can be made between osteoclastic and osteoplastic metastases. Both processes may be seen together in the same section, and attempts to grade the response of bone to tumor invasion[24] can be regarded as a pure intellectual curiosity. In osteoclastic metastases, the cancerous growth is accompanied by proliferation of connective tissue (Fig. 210), which may exceed that of the growths themselves. The margins of the tumor are surrounded by a wide zone of connective tissue proliferation, and the histologic appearance of this zone resembles that of osteitis fibrosa.

Osteoplastic metastases stimulate osteogenesis on the part of the local bone, and this is especially true in the metastatic, sclerosing lesions from the prostate. They are accompanied by an intense periosteal bone proliferation. The tumor cells may become enclosed in the bone, and bony tissue may be most abundant in very cellular areas of tumor cells. Extensive resorption of new bone by osteoclasts may follow the plastic process. There has been much speculation concerning the nature of the new bone formation in osteoplastic metastases, the number of theories being only

[24] Milch, R. A., and Changus, C. W.: Response of bone to tumor invasion, Cancer 9:340-351, 1956.

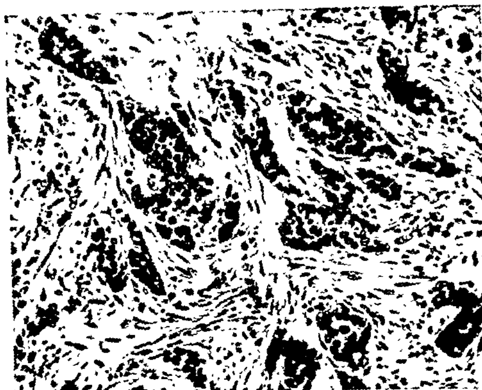


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[25] Willis, R A · The Spread of Tumours in the Human Body, St Louis, Mosby, 1952.

[26] Staley, C. J · Skeletal metastases in cancer of the breast, Surg, Gynec. & Obst. 102:683-688, 1956.

[27] Reynolds, M D, Lemon, H. M, and Byrnes, W W · Copper-resistant serum acid phosphatase. I. Method and values in health and disease. Cancer Res 16 943-950, 1956.

[28] Lemon, H M, and Reynolds, M. D : Regression of mammary cancer metastases following decline of serum acid phosphatase induced by anti-estrogen therapy, Proc. Am. A. Cancer Res. 2:129, 1956.

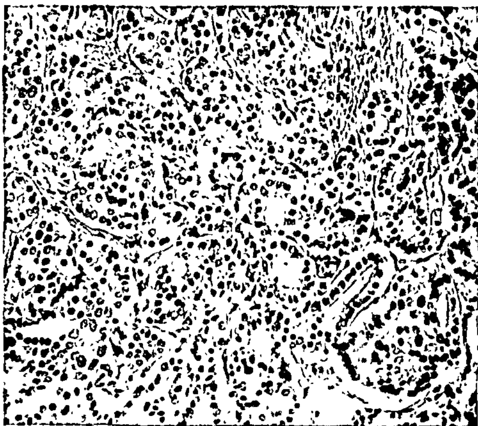




Fig. 211. (*Top*) histologic apearance of carcinoma of the prostate. (× 390) (*Bottom*) Extensive osteoplastic metastases of the lumbar vertebrae and upper third of the femur (primary in prostate). Note the pathologic fracture of the neck of the femur.

Carcinoma of the Prostate

Carcinoma of the prostate produces skeletal metastases in 40 to 50 per cent of cases. The bones most frequently affected are vertebrae. femur, pelvis, skull, ribs and sternum (Fig. 211). Clinically, metastatic carcinoma of the prostate is accompanied by progressive emaciation, severe anemia, and excruciating pains

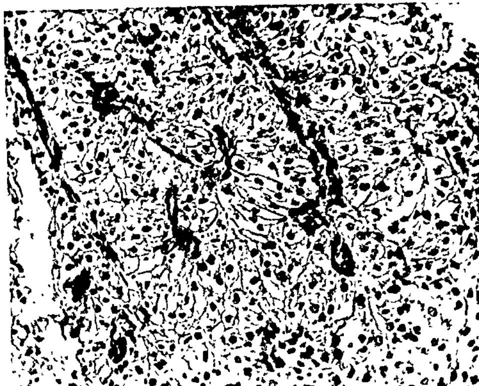


Fig. 212. Histologic appearance of a bone metastasis from a primary carcinoma of the kidney. (× 390)

in the affected bone. Roentgenologically, the lesions are of the osteoplastic type. There is a marked increase in the density of bone with light mottling which suggests some destruction. Such metastases are accompanied by an increase of the serum acid phosphatase. As in the case of metastatic mammary carcinoma, copper-resistant acid phosphatase has a better diagnostic and prognostic value than the usual acid phosphatase.[29] The presence of metastatic lesions is also associated with fibrinolysis. With fibrinolysis, there is a deficiency of

[29] Reynolds, M. D., Lemon, H. M., and Byrnes, W. W.: Copper-resistant serum acid phosphatase. I. Method and values in health and disease. Cancer Res. 16:943-950, 1956



Fig. 213. Metastatic carcinoma of the tibia (primary in the thyroid). (× 390)

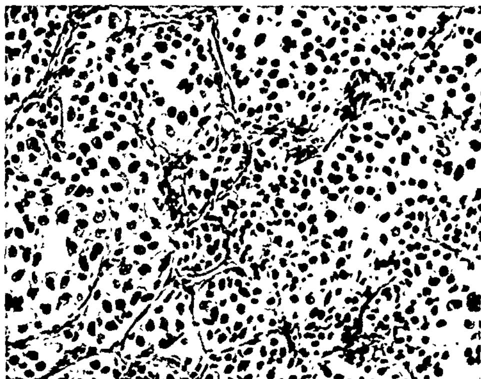


FIG. 214. Metastatic carcinoma of the spine (primary in the lung).

fibrinogen in the blood as well as an increase of the prothrombin time.[30]

Carcinoma of the Kidney

Bone metastases in carcinoma of the kidney occur in probably 30 to 50 per cent of the cases.[31] Bones most frequently involved are humerus, spine, femur, pelvis, ribs, bones of foot, skull and sternum. Pains and pathologic fractures are the main symptoms. In more than one half of the cases the secondary deposits are found as a single focus. In such cases it is imperative that the metastatic lesion be biopsied (Fig. 212) so that it will not be mistaken for a benign process or treated as a primary malignant bone tumor. Late bone metastases occurring up to 10 years after the removal of the primary growth are also recorded. The lesions are usually of the osteolytic type, and if the tumor extends through the bone into the soft tissue, pulsatile masses may be present.

Carcinoma of the Thyroid

Malignant thyroid tumors display a frequency of skeletal metastases of 25 to 30 per cent.[32, 33] The bones most frequently affected are the skull, ribs, the sternum, the spine and the humerus. The metastatic growths appear at epiphyses or along sutures. As in carcinoma of the kidney, carcinoma of the thyroid may develop only a single bone metastasis, sometimes before the primary growth is detected. Conversely, a single skeletal metastasis may appear many years after removal of the primary neoplasm. As a rule, the lesions are of the osteolytic type or of the cystic type, with encrusting bony shell.[34] Diagnosis is made by biopsy (Fig. 213). Radioactive iodine may be used in certain instances to confirm the diagnosis.[35]

Carcinoma of the Lung

Carcinoma of the lung metastasizes to the bones with a frequency of about 25 per cent of

30 Tagnon, H. J, Whitmore, W. F., Schulman, P, and Kravitz, S C · The significance of fibrinolysis occurring in patients with metastatic cancer of the prostate, Cancer 6 63-67, 1953

31 Geschickter, C. F., and Copeland, M. M Tumors of Bone, Philadelphia, Lippincott, 1949

32 Erhardt, O · Zur Anatomie und Klinik der Struma maligna, Beitr. klin. Chir. 35.343-464, 1902.

33 Dinsmore, R. S, and Hicken, N F.: Metastases from malignant tumors of thyroid, Am. J. Surg 24·202-224, 1934

34 Sherman, R. S, and Ivker, M.: The roentgen appearance of thyroid metastases in bone, Am J. Roentgenol 63·196-203, 1950

35 Lawrence, J. H., and Tobias, C. A.: Radioactive isotopes and nuclear radiations in the treatment of cancer, Cancer Res 16:185-193, 1956.



FIG. 215. Metastatic carcinoma of the tibia in a 9-year-old girl. (× 390) The primary tumor was found only at autopsy, and it was located in the lung.

cases. Both bronchogenic and alveolar cell carcinomas may give rise to skeletal metastases.[36] Such metastases are often accompanied by metastases to other organs. They are preferably located in the thoracic spine and in the ribs, less frequently in other bones. In Geschickter's series[37] the metastases involved the vertebral column in every case. Not infrequently, a small carcinoma of the lung may first reveal itself with a metastatic lesion. In such instances the microscopic pattern of the

[36] Nora, E., Baserga, R., and Saffiotti, U.: Il carcinoma bronchiolare, Arch. "de Vecchi" anat. pat. 20:1-29, 1953.

[37] Geschickter, C. F., and Copeland, M. M.: Tumors of Bone, Philadelphia, Lippincott, 1949.



FIG. 216. Metastatic carcinoma of the pelvic bones (primary in the pancreas). (× 390)

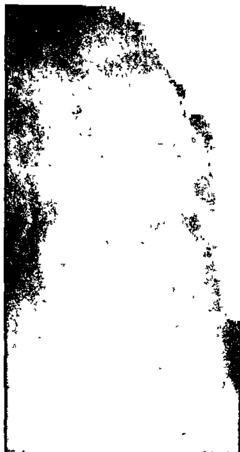


FIG. 217. Pathologic fracture of spine. Compression fractures of the vertebral body ordinarily are wedge-shaped, the apex being anterior, and the wedge smooth. In this case the maximum compression occurs centrally at a point of weakened resistance due to the neoplastic lesion. Such a fracture is often the first sign of metastases.



FIG 218. Carcinomatous metastases to the hip.

biopsy material will be the only available means to determine the diagnosis (Figs. 214, 215). The roentgenogram is characterized by osteolytic lesions.

Other Tumors

Skeletal metastases from other tumors are of minor clinical importance, because they are usually accompanied by widespread metastases in other organs. However, there are cases of carcinoma of the stomach in which the primary growth is a small symptomless one. The bones may be the principal or only site of metastases, and the patient's illness may closely simulate pernicious anemia.[38] Metastatic carcinoma from the rectum and the pancreas (Fig. 216) may simulate a primary bone lesion[39] Carcinoma of the liver[40, 41] and of the urinary bladder[42] metastasizes to the bone frequently. Carcinoma of the uterus metastasizes to the bones in 5 per cent of the cases, but the diagnosis is usually established before skeletal metastases appear. The skeleton, especially the skull with the typical brushlike appearance of the skull vault, is the favorite site of metastases from neuroblastomas. Other tumors metastasize less frequently, though in the literature are reported metastases from practically every primary growth, including carcinoma of the esophagus, the large intestine, the mouth or the pharynx, seminoma of the testis (particularly of the spine), retinoblastoma, chorionepithelioma, melanotic tu-

[38] Geschickter, C F., and Copeland, M M: Tumors of Bone, Philadelphia, Lippincott, 1949

[39] Bertin, E J.: Metastases to bone as the first symptom of cancer of the gastrointestinal tract; report of 3 cases, *Am J Roentgenol 51.614-621, 1944*

[40] Auerbach, O, and Trubowitz, S · Primary carcinoma of the liver, with extensive skeletal metastases and panmyelophthisis, Cancer 3 837-843, 1950.

[41] Edmondson, H A, and Steiner, P. E. Primary carcinoma of the liver, Cancer 7 462-503, 1954

[42] Kretschmer, H. L. Carcinoma of the bladder with bone metastases, Surg, Gynec & Obst. 34 241-246, 1922

mors,[43] carcinoma of the ovary, retroperitoneal carcinoma, salivary mixed tumor, carcinoma of the ureter, Ewing's sarcoma,[44] and even osteogenic sarcomas.[45]

Sarcomas of soft tissue origin do not as frequently involve bone. So-called Hodgkin's disease frequently implicates bone but only in advanced cases. Lymphosarcomas may involve bone, usually as an osteolytic process, but invariably the lymphosarcoma has been noted previously and diagnosed. Metastases of many lymphosarcomas into bones, especially the small lymphocytic, but also forms of reticulum cell sarcomas, do not cause appreciable destruction of the bone structure but markedly replace the basic marrow tissue.[46]

TREATMENT

Metastatic growths in the skeleton are nearly always multiple (156 out of our 176 cases). The suggestion of some surgeons that a "solitary" metastasis might justify amputation of a limb must be regarded as applicable only in the rarest cases.

Bone metastases, especially a single osteolytic one, may respond favorably to x-ray treatment. The more extensive the metastases, the poorer is the response to treatment. The dosage should not be pushed to the point where it induces radiation necrosis or enhances the likelihood of pathologic fracture. In general, the results, while distinctly worthwhile in suitable cases, are on the whole unpredictable.[47]

Radioactive isotopes and nuclear radiations have also been used in the treatment of bone metastases Radioactive iodone (I^{131}) has been used in the treatment of thyroid metastases.[48] By the use of the whole body scintillation scanner, developed by Anger and Tobias for photographing gamma-ray-emitting isotopes in the body, the I^{131} uptake by the metastases can be located and treatment given at the same time. Uptake in metastases can be stimulated by surgical thyroidectomy or by the administration of propylthiouracil and thyroid-stimulating hormones.[49]

However, the radiation dose may not be uniformly distributed in the tumor tissue, and surgery and x-rays continue to be the primary tools for treating thyroid cancer. Radioactive phosphorous (P^{32}) has been used in the treatment of metastatic carcinoma of the mammary gland and prostate cancer metastatic to bone. The results are contradictory, but there is some relief of pain and occasional evidence of healing. This type of therapy may have a small place in the palliation of metastatic cancer. The cobalt bomb teletherapeutic units,[50] which provide a gamma ray source and have certain advantages over x-ray equipment, can also be used in the treatment of bone metastases, but one cannot expect much improvement in the end results over the conventional methods which use x-ray or other gamma ray sources.

Hormonal treatment has been of considerable value in the treatment of metastatic bone cancer, especially in carcinoma of the prostate and in carcinoma of the mammary gland.[51] The endocrine treatment of prostatic cancer consists of anti-androgenic measures; both orchiectomy and phenolic estrogens are effective in controlling cancer of the prostate in certain cases. There is no doubt that patients with prostatic cancer who respond favorably to castration and/or estrogen therapy live more comfortably and longer than patients not treated by these methods. In cases that do not respond to castration and/or estrogen therapy, bilateral adrenalectomy has been carried out Remissions following adrenalectomy have been reported, but it is now known

[43] Selby, H M, Sherman, R. S, and Pack, G. T.: A roentgen study of bone metastases from melanoma, Radiology 67.224-228, 1956.

[44] Hodges, P. C, Phemister, D B, and Brunschwig, A The roentgen-ray diagnosis of diseases of bone *in* Ross Golden's Diagnostic Roentgenology, New York, Nelson, 1941

[45] Dresser, R, and Dumas, C. Osteogenic sarcoma, Am J Roentgenol 23 65-66, 1930

[46] Hirsch, E F · Pathology in Surgery, Baltimore, Williams & Wilkins, 1953

[47] Lichtenstein, L: Bone Tumors, St. Louis, Mosby, 1952

[48] Lawrence, J. H, and Tobias, C. A.. Radioactive isotopes and nuclear radiations in the treatment of cancer, Cancer Res. 16.185-193, 1956

[49] Seidlin, S M, Rossman, I, Oshry, E, and Siegel, E · Radioiodine therapy of metastases from carcinoma of the thyroid. a six-year progress report, J Clin. Endocrinol. 9.1122-1137, 1949

[50] Lawrence, J H, and Tobias, C. A: Radioactive isotopes and nuclear radiations in the treatment of cancer, Cancer Res 16.185-193, 1956

[51] Huggins, C: Control of cancers of man by endocrinologic methods; a review, Cancer Res. 16 825-830, 1956.

that adrenalectomy benefits only a small percentage of patients with prostatic cancer in relapses after orchiectomy and the administration of estrogens.

The endocrine treatment of cancer of the mammary gland consists essentially of ovariectomy. The regression of mammary cancer can be profound, remissions occurring in approximately 30 per cent of mammary cancers. Bilateral adrenalectomy has also induced profound and prolonged regressions of disseminated mammary cancer. In a proportion of cases both subjective and objective improvement have been achieved. Maintained adequately with steroid substitution therapy, the adrenalectomized patients have a healthy appearance, are not incapacitated and are able to engage in all of their usual activities. In postmenopausal women, adrenalectomy frequently is followed by a recrudescence of menopausal symptoms which had been dormant for some years. At least in some instances, substitution therapy with cortisone may have a favoring action on the growth of metastases.[52] Another endocrine approach to the therapy of metastatic breast cancer is hypophysectomy, which can be accomplished either by surgery or by external radiation with a beam of protons derived from the cyclotron.[53]

Testosterone has long been known to delay recurrence following radical mastectomy for cancer of the breast, and it can produce temporary improvement in women with metastatic mammary cancer.[54] However, exacerbation of metastatic mammary cancer in bone has been observed during treatment with androgens. The accelerated tumor growth was accompanied by hypercalcemia and/or hypercalciuria. The stimulatory effect was related to the possible conversion of testosterone to estrone or estradiol.[55]

[52] Baserga, R., and Shubik, P.: Action of cortisone on disseminated tumor cells after removal of the primary growth, Science 121:100-101, 1955.

[53] Huggins, C.: Control of cancers of man by endocrinologic methods; a review, Cancer Res. 16 825-830, 1956.

[54] Loeser, A A.: Mammary carcinoma response to implantation of male hormone and progesterone, Lancet 2.698-700, 1941.

[55] Myers, W. P L., West, C. D., Pearson, O. H., and Karnofski, D A : Androgen-induced exacerbation of breast cancer measured by calcium excretion; conversion of androgen to estrogen as a possible underlying mechanism, J.A.M.A. 161:127-131, 1956

15

Diseases of Muscle

HISTOPATHOLOGY OF SKELETAL MUSCLE[1,2]

Skeletal muscle is subject to *degeneration* (due to infection or trauma) and *necrosis* (due to ischemia or trauma). The ensuing process is similar to that in other tissues. First, the debris is removed by mononuclear cells and macrophages. Then repair is effected (1) by regeneration of muscle from surrounding normal muscle fibers, and (2) by ingrowth of granulation tissue and fibrosis. Regardless of the mode of destruction, whether by trauma, ischemia or toxic degeneration, regeneration of muscle can occur.

Microscopic Appearance. Necrosed muscle fibers undergo fragmentation, stain irregularly, and the sarcolemmal nuclei become shrunken and pyknotic. The surrounding undamaged muscle is prevented from retracting by the connective tissue. Within a few days, mononuclear inflammatory cells and macrophages invade the necrotic zone and remove the debris. After about 4 days, surrounding muscle fibers send deeply basophilic, multinucleated, protoplasmic processes into the area. Granulation tissue invades at the same time. After 1 week, the slender straps are prolific and penetrate and close gaps quickly, especially where sarcolemmal tubes have persisted. When sarcolemmal tubes are absent, ingrowth of multinucleated sarcoplasm is delayed, and fibroblastic replacement is pronounced. After 2 weeks, the new fibers acquire sarcolemmal sheaths, cross striations appear, the fibers enlarge, basophilic staining is lost, and a mature appearance is acquired. The new muscle fibers are irregularly woven together. After 6 weeks, the area grossly and microscopically appears normal except for occasional small areas of fibrosis.

This sequence of events indicates that damaged muscle should be protected for a minimum of 6 weeks before allowing activity.

Zenker's Degeneration or Waxy Degeneration. This is a hyaline (glasslike) degeneration occurring in skeletal muscle. In a severe form, it is found in severe infections, such as pneumonia, and is observed best in the diaphragm and the rectus abdominis. Otherwise, during an acute infectious disease, it occurs to some degree in all skeletal muscle. Microscopically, the muscle fibers are swollen (due to increased osmotic pressure), homogeneous without striations, and stain intensely acidophilic. Clinically, such muscle is easily ruptured, causing intramuscular hemorrhage. The importance of restricting activity after a severe febrile illness is readily apparent.

Histology of Denervated Muscle. The muscle undergoes progressive atrophy. In each muscle bundle the peripheral fibers in particular diminish in caliber, although sarcolemmal nuclei, longitudinal myofibrils and cross striations persist during the first 3 weeks. However, by 3 weeks wallerian degeneration is complete, and fibrosis replaces the muscle fibers. When a denervated muscle is traumatized, it regenerates exactly as the normal muscle does.

Identification of Striated Muscle Tumors. Certain neoplasms which stem from skeletal muscle present such a pleomorphic appearance that their origin is difficult to ascertain. Strands of acidophilic tissue are suspected of being muscle fibers. Their cross striations can be displayed best by staining with phosphotungstic acid.

[1] Clark, W E Le Gros: An experimental study of regeneration of mammalian striped muscle, J. Anat. 80:24, 1946.

[2] Saunders, J. H, and Sissons, H A: Effect of denervation on regeneration of skeletal muscle after injury, J Bone & Joint Surg 35B 113, 1953

TUMORS OF SKELETAL MUSCLE[3]

Rhabdomyoma

This benign tumor arising in striated muscle is extremely rare. It tends to form an infiltrative, nonencapsulated nodule.

Microscopic Appearance. The tumor is *composed chiefly of interlacing bundles of* long straplike cells with single nuclei, myofibrils and occasional cross striations. The cytoplasm is acidophilic but not granular. No mitoses are seen.

Rhabdomyosarcoma

This rare malignant tumor can occur at any age, but the greatest incidence is between 30 and 60 It is usually located in the voluntary muscle of the thigh, the buttock or the shoulder girdle. It forms a palpable soft mass which is painless except when it compresses a nerve. It may break through the skin as a red, fungating mass, especially after incomplete local excision. It generally has a deep-red, meaty appearance and is surrounded by a pseudocapsule It can invade and destroy adjacent bone

Microscopic Appearance. A wide variety of pleomorphic cells are observed. The nuclei are large, the cytoplasm deeply acidophilic, and many cells contain cross striations characteristic of skeletal muscle. Four types of cells can be identified:

1. *Strap Cell.* The most common type. Long, meandering cytoplasmic process containing two or more nuclei ("tandem nuclei")
2. *Racket Cell* The nucleus bulges at one end, and the tapering tail of cytoplasm resembles the handle of a tennis racket.
3. *Tumor Giant Cell.* Cytoplasm vacuolated resembles a spider web. Nuclei are extremely large and anaplastic. Vacuoles *contain* glycogen.
4. *Embryonal Myoblast.* Small size, dense nuclei, scanty cytoplasm.

CAVERNOUS HEMANGIOMA OF STRIATED MUSCLE[4,5,6]

This is a not uncommon benign tumor composed of many thin-walled blood vessel spaces within a densely fibrous tissue. The cause is unknown. Young individuals are affected most often. The majority of hemangiomas are located intramuscularly, and usually a single tumor is confined to a single muscle. The quadriceps is involved most frequently. Rarely, the cavernous *hemangioma involves a group* of muscles or forms part of extensive angiomatosis of an entire extremity.

Pathology

Grossly, the muscle appears swollen, and its color and consistency vary, depending on the relative proportion of cavernous sinuses to fibrous tissue replacing the muscle substance. On cut section, the muscle locally contains dense interlacing fibrous strands extending in *all directions and gradually merging* with surrounding normal muscle fibers. The fibrous strands enclose numerous thin-walled bleeding sinuses, many of which contain thrombi. Characteristically, the tumor contains multiple minute, grayish-white, round, hard nodules representing calcified thrombi.

Histologically, a large *number* of vascular spaces lined with a single layer of endothelium are observed. Surrounding these is a dense fibrous matrix within which may be seen degenerating muscle fibers. The walls of arterioles display fibrous thickening. The cavernous spaces contain thrombi in various stages of organization and calcification.

Clinical Picture

A sensation of tenseness or actual pain, *particularly on contraction of the muscle, is the* usual complaint. The muscle appears diffusely swollen. On palpation, a poorly delimited mass is perceived. It is tender, and its consistency varies from soft to hard, depending on the relative amounts of vascular spaces to fibrous stroma. Multiple pea-sized nodules are often palpable. The tumor grows slowly. It is often confined to one muscle, commonly the quadriceps, and it moves and becomes prominent with contraction of the muscle. The mass may

[3] Soule, E. H: Am. Acad Orthop. Surgeons, Lect 14 321, 1957.

[4] Jones, K. G.: Cavernous hemangioma of striated muscle; a review of the literature, J. Bone & Joint Surg. 35A:717, 1953.

[5] Selakovitch, W. G., and Sherman, M. S. Hemangioma of the musculoskeletal system, Ochsner Clinic Rep 2 41, 1956

[6] Bendeck, T E., and Lichtenberg, F.: Cavernous hemangioma, Ann. Surg. 146:1011, 1957.

be rendered tense and more easily palpated by applying a tourniquet, compressing the veins above it. On releasing the tourniquet, the swelling once again becomes soft and boggy. Elevation and dependency of the extremity produce similar changes. Aspiration yields blood.

Roentgenologic Findings

A soft tissue density is observed within the swollen muscle. Opacities of phleboliths lie within the tumor shadow. These are differentiated from similar opacities of trichinosis, which are much smaller, and cysticerci, which are elongated and less dense. Periosteal reactive ossification is observed in adjacent bone.

Complications

Rupture of the muscle has been observed.

Treatment

Complete surgical excision is advisable. Other more conservative therapeutic measures are uncertain and often are followed by recurrence. If the defect created by removal is large, fascial replacement or muscle transfer may be necessary.

TRAUMATIC CONDITIONS OF MUSCLE

Muscle can be injured by direct trauma, such as a forceful blow, or indirect trauma, which stretches and tears the muscle fibers.

Direct Trauma

Crushing of nerve fibers and connective tissue sheaths causes a variable amount of necrosis which to a great extent is proportionate to the degree of vascular damage. The biceps and the anterior tibial muscles are especially prone to extensive post-traumatic necrosis because of a solitary blood supply. When the traumatic force is applied to large areas of muscle, parenchymal necrosis and subsequent scar tissue replacement is extensive. A muscle injury localized to a small area can be followed by complete muscle regeneration with a minimum of scar.

Muscle Rupture and Fascial Tears

The mechanism of indirect injury is either (1) prolonged exercise in an untrained individual, or (2) a violent contraction of certain muscles. Muscle fibers rupture, hemorrhage and edema follow, and the muscle herniates through a rent in the overlying fascia or epimysium. A soft elastic tumor which is not adherent to the skin presents itself. It becomes tense and painful on contraction of the muscle. It can be reduced by compression while the muscle itself is relaxed. The biceps, the rectus femoris and the gastrocnemius are involved most often. With passage of time, the hernia becomes fibrosed. Rarely does it necrose.

Complete rupture of a muscle takes place most often through the muscle belly, less often at the musculotendinous or tendo-osseous junctions. Its occurrence in certain muscles is favored by muscle action peculiar to certain occupations. For example, the biceps and the triceps are frequently ruptured in baseball pitchers; the calf muscles, in boxers; the thigh adductors, in horseback riders, etc. Rupture may occur also in severe tetanus, or in electric shock injuries, which cause violent concurrent contractions of antagonist muscles. Clinically, the sequence of events are: (1) an audible snap, associated with severe pain, occurring during a strong contraction; (2) swelling or bulge of retracted muscle; (3) weakness or paralysis. A partial muscle rupture exhibits only localized pain and tenderness and is often diagnosed as a sprain or a "charleyhorse." The degree of weakening of muscle power is proportionate to the extent of disruption. Often an incomplete rupture may become complete the following day.

Pathologic rupture may occur in certain conditions characterized by parenchymatous degeneration, e.g., trichinosis, typhoid fever, tuberculosis and other infections.

Muscle Hemorrhage

Hemorrhage occurs in muscle from several causes:

1. **Trauma.** The exposed limb muscles are predisposed.

2. **Severe Infectious Diseases.** Generalized Zenker's waxy degeneration of sarcoplasm renders all muscle quite fragile. A thin exposed muscle, such as the rectus abdominis, is predisposed. Streaks of extravasated blood develop between the muscle fibers, or a discrete hematoma may form. The muscle is diffusely tender and painful.



FIG. 219. Heterotopic ossification.

3. Hemorrhagic Diseases. Scurvy and thrombocytopenic purpura are examples.

If hemorrhage is small, it may be absorbed. If large, the clot may become encapsulated or organized or converted to cartilage or bone Muscle fibers about a hemorrhage may be destroyed or undergo hyaline degeneration. Later, regeneration occurs by accumulation of sarcoplasm about multiplying nuclei, forming the giant cells characteristic of muscle reconstruction.

TRAUMATIC MYOSITIS OSSIFICANS

Synonyms: Traumatic ossifying myositis; ossifying hematoma, calcified hematoma; traumatic parosteal bone formation.

This condition may be defined as the formation of bone in muscle as the result of injury. It may follow a simple severe blow or a series of repeated minor traumata (myositis ossificans circumscripta).

Theoretically, this is supposed to be due to displacement of periosteum which continues its osteogenic function in a heterotopic location. A hematoma seems to be a necessary prerequisite. In the process of resorption and organization of the blood clot by invading granulation tissue, the site becomes edematous and apparently a suitable medium for formation of osseous tissue. The displaced primitive osteogenic tissue forms osteoid trabeculae directly in fibrous tissue comparable with membranous ossification. Only rarely does cartilage form and is converted to bone by direct metaplasia or by endochondral ossification. The irregular cancellous type of bone is resorbed and replaced by more dense and laminated bone as a final stage. This pathologic sequence explains the x-ray picture of early indefinite opacity in the soft tissue adjacent to the main bone and separated from it by a normal soft-tissue interval. The shadow increases in size and density over a period of 3 to 6 weeks, becomes well outlined, then recedes a little to assume its permanent size and configuration. It is usually elongate, parallel with the bone and occasionally attached to it, contains bony laminations, may have the density of the adjacent bone and is sharply outlined. This is the stage of "maturity."

Histologically, the picture varies, depending on the stage of progression. Bony trabeculae irregularly arranged are surrounded by many osteoblasts and are embedded in an adult or edematous fibrous tissue. Prior to this stage, mainly imperfect osteoid tissue is found. A later stage will show large segments of laminated bone. Occasionally, the stroma may be quite vascular.

Clinical Picture. The condition occurs as a result of a single or repeated trauma. The brachialis anticus is a favorite site after a posterior dislocation of the elbow. During development of the extra-osseous mass, the elbow area is quite swollen and tender, and active and passive motion is greatly restricted. Gradually, as the pain and the general swelling are reduced, a circumscribed indurated, later hard, tumor mass is palpable over the anterior aspect of the elbow. Active extension of the joint is limited by virtue of inelasticity of the muscle, and flexion is prevented by obstruction offered by the mass.

Ossification in the deltoid is common in foot soldiers due to the trauma of carrying a rifle. The constant pressure of the saddle against

the adductors in riders causes ossification in that muscle. The syndrome is known as "Prussian disease."

Subperiosteal hemorrhage followed by formation of bone may be included in this category, although not occurring in muscle. The pathogenesis is similar. The ossification in the quadriceps area occurring in football players is an example.

Treatment. The growth should not be removed in the premature stage. The result of such ill-advised attempts is disastrous. The ossification becomes exuberant, infiltrates beyond the original site and compromises the soft tissues about the joint beyond hope of repair. Restraint is the watchword. When by serial roentgenograms the mass is dense, well delineated and at a standstill, it may safely be removed. One may ensure against recurrence by waiting until a very late date. It may be possible to prevent myositis ossificans by aspirating the original hematoma.

MYOSITIS OSSIFICANS PROGRESSIVA

This congenital condition starts without antecedent trauma before or shortly after birth. It consists of frequently repeated episodes of sudden extension of ossification in the muscles, the fasciae, the tendons and the aponeuroses. The only laboratory finding is an elevated eosinophil count. The blood chemistry is normal. Other congenital deformities as microdactyly, brachydactyly, Klippel-Feil syndrome, etc, are frequently associated. The ossification usually starts in the upper back muscles, particularly the trapezius and the latissimus dorsi, and spreads distalward and eventually involves soft-tissue structures throughout the body. The interphalangeal joints of the thumb, the large toe and the spine are liable to fuse. All joint motion is finally lost, and the patient succumbs to intercurrent infection. The condition is very rare

Treatment

There is no known effective treatment The administration of corticotropin seems to have a deterrent effect on the heterotopic bone formation. The eosinophil count drops, and joint motion may even increase[7]

[7] Lockhart, J D, and Burke, F G Myositis ossificans progressiva, A M A Am J Dis. Child 87:626, 1954

ANTERIOR TIBIAL SYNDROME

(Traumatic Necrosis of Pretibial Muscles)

This condition is characterized by necrosis of the group of muscles within the anterior tibial compartment.

Clinical Picture

Following a period of unaccustomed vigorous and prolonged exercise, the patient, often a young adult male, develops aching pain in the anterior aspect of one or both legs. A firm, tender swelling is palpated over the entire anterior tibial compartment. Paralysis of the pretibial muscles (anterior tibial, extensor hallucis longus, extensor digitorum longus) develops quickly, the resultant drop foot being less pronounced than that usually associated with nerve lesions. The affected muscles do not respond to either faradic or galvanic current, and electromyography shows complete absence of electric activity. Passive movements of these muscles are painful. The overlying skin is erythematous, glossy, edematous and warm. Mild systemic symptoms as a low-grade fever, leukocytosis and albuminuria are not uncommon. Rarely, implication of the anterior tibial nerve is revealed by paralysis of the extensor digitorum brevis and an area of sensory loss on the dorsum of the foot between the first and the second toes.

The pain, dermal erythema and systemic symptoms subside within a few days. However, the paralysis may persist. Fibrous contracture develops and resists plantar flexion of the foot.

Pathology

Grossly, when the anterior crural fascia is incised, the swollen muscles bulge into the wound. During the acute stage the muscles are soft, friable and reddish gray. Later they are fibrotic, indurated and grayish white.

Microscopically, the muscle fibers exhibit degeneration by swelling, intense acidophilic staining, loss of nuclei and striations, shrinking and pyknosis of nuclei, and granular and vacuolar appearance. Red blood corpuscles infiltrate between the fibers Macrophages engulf and remove the degenerated acidophilic material and hemosiderin. Regenerative activity is evident in large numbers of newly formed nuclei surrounded by sarcoplasm form-

ing giant cells. Fibrous tissue proliferation is extensive and replaces destroyed muscle fibers.

Pathogenesis

The actual cause is unknown. The clinical and pathologic picture suggests that a muscle which is not yet adapted to vigorous contractions produces excess quantities of metabolites which causes it to swell. Swelling of the pretibial muscles increases the pressure within their rigid unyielding fascial compartment, resulting in ischemic necrosis.

Treatment

Immediate incision of the crural fascia will liberate the swollen muscles. Foot and ankle motion must be restricted for several weeks until muscle regeneration and fibrous replacement have occurred If necrosis is not too extensive, regeneration with recovery of muscle function may occur. During the acute stage, manipulation of the paralyzed legs must be avoided, because sudden severe destruction of muscle fibers may lead to myoglobinuria, lower nephron nephrosis and death.[8]

"Shin Splints"

This term is applied to a common condition of pain and tenderness over the anterior tibial muscles occurring at the beginning of an athletic training program. The affected muscles are swollen but not paralyzed. Active or passive attempts at movement aggravate the pain. It is entirely possible that "shin splints" is a mild form of the anterior tibial syndrome.

The treatment is rest and lukewarm heat applications. Subsequent exercises should be instituted slowly and gradually.

ISCHEMIC NECROSIS OF MUSCLE

Muscle tissue has such rich collateral circulation that infarction is rare in the absence of peripheral circulatory diseases. The causes of ischemic necrosis of muscle are (1) occlusion of a main artery by embolism, thrombosis or compression; (2) thrombosis of multiple peripheral vessels as in Buerger's disease or atherosclerosis; (3) thrombosis of multiple intramuscular vessels as in polyarteritis nodosa; (4) swelling or hemorrhage in certain muscle groups enclosed within rigid fascial compartments, e.g., the pretibial muscles.

Pathology

Necrotic muscle is friable with patches of yellowish-green discoloration. The margins of the infarct where the circulation is preserved are a mottled red and yellow; rose-colored foci appear later. Yellow coloration represents destroyed muscle; rose color, the regenerating muscle.

Microscopically, necrotic fibers are fragmented and stain irregularly. Sarcolemmal nuclei are shrunken and pyknotic. At the margins of the necrotic zone, fibers undergo degenerative changes such as waxy or hyaline appearance, loss of striations, and pyknosis and disappearance of sarcolemmal nuclei. Next, neutrophilic leukocytes and macrophages infiltrate and remove the damaged fibers. Adjacent to the infarct new fibers are formed. These young fibers are distinguished from old ones by their small size and basophilic coloration in H-E stains. Finally, vascularized connective tissue replaces the disappearing muscle fibers.

Massive necrosis may not occur, particularly when reduction of circulation is gradual. Instead, there develops a diffuse interstitial fibrosis or multiple foci of necrosis with replacement fibrosis. Under the microscope, interstitial fibrosis is seen as individual healthy muscle fibers or fascicles separated by connective tissue. Scattered collections of lymphocytes and histiocytes infiltrate the perimysium and surround blood vessels. The process suggests vascular congestion and edema causing diffuse alteration of muscle and hyperplasia of connective tissue.

Massive necrosis of an entire muscle is inevitable in the case of certain muscles whose single nutrient artery is compromised by thrombosis, embolism or injury. Muscles with this peculiarity include the gastrocnemius, the long head of the biceps and the anterior tibial muscles.

Multiple scattered infarcts throughout a muscle are characteristic of periarteritis nodosa, which causes thrombosis of many minute intramuscular vessels. On the other hand, atherosclerosis or the necrotizing arteriolitis of

[8] Robinson, G L: Scattered muscle necrosis associated with post-traumatic uremia, Lancet 1 799, 1950.

malignant hypertension rarely occurs in vessels as small as those found in muscle.

Experimental evidence seems to indicate that venous occlusion is more likely to produce a diffuse interstitial fibrosis, whereas arterial occlusion may have no effect whatsoever or may cause multiple foci of necrosis or massive necrosis.[9]

Volkmann's Ischemic Contracture[10]

This is a progressive post-traumatic muscle contracture, a result of ischemic necrosis. It occurs most commonly in the forearm muscles as a result of trauma to the brachial vessels at the elbow. Serious crippling of hand function results. (This condition is discussed in the section on Circulatory Diseases.)

MYOSITIS

The term "myositis" is defined as inflammation of muscle. This implies that an irritative agent, such as bacteria, parasites, virus, etc., produces a reactive vasodilatation, outpouring of serum and cells, and healing by granulation tissue. The inflammatory process takes place in the interstitial tissue, whereas the muscle fibers are destroyed by toxins, compression of accumulating exudate, or ischemia secondary to focal thrombosis. Subsidence of inflammation is followed by attempted regeneration of muscle fibers. True inflammatory myositis should be distinguished from parenchymatous degeneration which occurs in the course of a systemic infectious disease and displays no inflammatory phenomena.

Bacterial Myositis

Acute Suppurative Myositis. Abscess formation in muscle is rare. It usually arises by spread from an adjacent focus, such as osteomyelitis, or by a puncture wound; rarely, it is hematogenous. The common causative organisms are the staphylococcus and the streptococcus.

Pathology. Early, edema and cellular infiltration occur in the interstitial tissue. Muscle fibers are intact but display parenchymatous changes as swelling, obscuring of cross-striations, and granularity or vacuolization. The process may subside with complete restoration of normal histology. It may extend and cause a diffuse interstitial cellular infiltration with vascular thromboses, producing an indurated phlegmonous mass of tissue. An exudate accumulates from continued outpouring of cells, serum, and liquefaction of necrotic tissue. Muscle fibers are destroyed by compression, toxic degeneration, ischemic necrosis and liquefaction. When the muscle is enclosed in a rigid fascial compartment, the intrafascial tension mounts rapidly, and destruction of muscle fibers develops quickly. Abscesses may become encapsulated by fibroblastic granulation tissue and, after resolution or evacuation, are transformed into fibrous tissue. Extensive destruction of muscle is followed by replacement by dense cicatrix leading to contracture.

Clinical Picture. Constitutional symptoms of chills, fever and sweats are followed shortly by local pain in the affected muscle. Initially, the overlying skin may be reddened and warm. The muscle is swollen, indurated and very tender, but these findings may be masked when the muscle, such as the anterior tibial, is enclosed in a rigid fascial compartment. Softening and fluctuation may become apparent, and spontaneous drainage may occur. At first, muscle power is reduced, and electric excitability is diminished, but reaction of degeneration never does develop. As destruction progresses, loss of muscle function is complete.

Healing in the early stage permits complete restoration of muscle function and power. A slight amount of residual fibrosis may be manifest only in easier fatigue following extreme activity demanding strong muscular contractions. Excessive fibrosis produces contractural deformity. Severe muscle tissue loss with minimal fibrous replacement in effect produces permanent paralysis and deformity due to unopposed pull of the antagonist.

Treatment. Diagnosis and treatment are urgent. A small amount of exudate, which under other circumstances is relatively benign, *can effect rapidly* mounting tension within a fascial compartment enough to destroy the entire muscle. Immediate incision and drainage, even before signs of fluctuation, are mandatory. Antibiotics of *several kinds* are given

[9] Brooks, B: Pathological stages in muscle as a result of disturbances of circulation. An experimental study of Volkmann's ischemic paralysis, Arch. Surg. *5:188, 1922.*

[10] Volkmann, R, von: Krankenheiten der Bewegungsorgane, Handbuch der Chirurgie 2:846, 1872.

while awaiting bacterial culture and sensitivity tests. Hot, moist compresses are applied, and blood transfusions are given The extremity is elevated and splinted in the position of function to counteract contracture and provide rest to the muscle.

Clostridial Myositis
(Anaerobic Myositis)

Gas gangrene is caused by saprophytic bacteria, especially *Clostridium welchii.* Healthy muscle is ordinarily resistant to clostridial organisms, but trauma renders it susceptible. The tissue is rapidly decomposed with formation of gas bubbles (carbon dioxide from muscle glycogen) and edema.

Clinical Picture. Gas gangrene usually follows a deep puncture wound, as is common with compound fractures. The onset is acute with an abrupt fever, chills and prostration. Pulse and respiratory rates rise, blood pressure falls. The wound is severely painful, swollen and red. The surrounding skin is grossly edematous and red at first, then develops a bronze or copper-colored appearance. Serosanguineous exudate drains profusely from the wound. The swollen tissues may mask the crepitus of gas bubble accumulations, which gradually spreads in the subcutaneous tissues along fascial planes. A characteristic acid odor is noted Rarely, septicemia may develop with jaundice, hemoglobinemia, hemoglobinuria and death.

Pathology. The muscle appears red, friable and, in places, semitranslucent. Microscopically, all tissue elements (endomysial connective tissues, sarcolemma, vascular, neural) about the wound are necrotic. The muscle fibers undergo coagulation necrosis with loss of striations, and nuclei are pale staining or disappear. Dead fibers become thin and disappear or they fuse and liquefy. Large numbers of gram-positive bacilli are prominent in both the muscle fibers and the interstitial tissues. Exudation of fibrin is profuse Beyond the necrotic zone, newly involved tissue displays intense congestion, hemorrhages, leukocytic infiltration, edema separating muscle fibers, obscuring of striations, pyknosis and pallor of sarcolemmal nuclei, and granular destruction of interstitial connective tissue.

If infection is controlled, the necrotic muscle is removed, muscle regeneration is sporadic and inadequate, and fibrosis with marked diminution of muscle size is the usual result.

Treatment. Prophylaxis by gas gangrene antiserum is of doubtful value except for prolonging the incubation period.

Surgically, the infected area is opened widely because the bacteria cannot survive in the presence of oxygen. Excision of infected tissues must be radical. Peroxide of hydrogen or zinc is instilled in the wound. Blood transfusions are most important. Polyvalent antitoxin is administered intravenously in doses of 100,000 I.U. Antibiotics, as penicillin and the tetracyclines, are given locally and systemically. If amputation becomes necessary as a lifesaving procedure, it should be of the open type, to be closed later when infection has been overcome.

Tuberculous Myositis

Striated muscle is never involved by a primary tuberculous lesion except, rarely, by accidental inoculation with an infected needle. More common forms are the following:

1. **Extension From Neighboring Focus.** A cold abscess erodes the epimysium and spreads along the sheaths. Muscle fibers and connective tissue are destroyed by caseation necrosis. An example is the common psoas abscess.

2. **Hematogeneous Spread (Miliary T. B.).** The lesion is usually small and confined to one muscle. It forms an abscess and a sinus tract, or a nodular sclerosis with eventual calcifications. Tubercles form in the interstitial connective tissue in relation to intermuscular vessels. Muscle fibers adjacent to the tubercle are destroyed. Only when an abscess forms do muscle fibers at a distance undergo pressure atrophy. Damaged muscle fibers may regenerate. Muscle lesions, as a rule, are asymptomatic.

3. **Polymyositis.** Multiple involvement with tubercles (which may be difficult to differentiate from Boeck's sarcoid) are manifested clinically by progressive weakness, atrophy, reflex loss and pronounced contractures, causing severe disability. Microscopically, muscle fibers undergo granular or fatty degeneration and are replaced by fibroblastic connective tissue.

Syphilitic Myositis

This is rare. The lesion consists of a gumma, a solitary circumscribed focus of yellowish-gray tissue enveloped by dense connective tissue. The sternocleidomastoid and the biceps are involved most often. Another type of involvement is a diffuse interstitial myositis in which large amounts of fibrous connective tissue replace degenerated muscle fibers in many muscles. These muscles are unusually hard and painless. Contractures are frequent.

Parasitic Myositis

Trichinosis is the most frequent parasitic infection in muscle. About 17 per cent of the adult population in the United States is said to be infected. Other parasites include *Cysticercus cellulosae*, the larval parasite of the pork tapeworm; *Taenia solium*, the embryo parasite of the tapeworm *Echinococcus granulosis;* the *Sarcosporidia* parasite, which occurs in the striped muscle of most domestic animals; *Toxoplasma;* and *Trypanosoma.* From the orthopaedic standpoint, trichinosis is most important, as it produces muscle symptoms, findings and sequelae. The tendency of other parasites to localize in skeletal muscle is without symptoms, but muscle biopsy is often used for diagnosis.

Trichinosis. The cause is *Trichinella spiralis*, a nematode infecting man by ingestion of raw or poorly cooked pork. The encysted trichinae escape in the intestinal tract as the pork muscle is digested, and the larvae penetrate the duodenal mucosa. The fertilized female deposits batches of embryos which enter the lymphatics and emigrate through the blood stream to various tissues. Only in striated muscle are conditions favorable for their growth.

Clinical Picture. Three stages are recognized:

1. *Intestinal Stage.* This is characterized by symptoms of gastroenteritis during the first week.

2. *Stage of Muscular Invasion.* This lasts about 5 weeks. Symptoms include chilliness, irregular low-grade fever, pains in the trunk and the extremities, muscular tenderness, edema of the conjunctivae and the periorbital tissues, fatigue and, rarely, prostration. Muscle weakness may appear and when severe may be associated with loss of tendon reflexes. Muscle weakness may be generalized or it may be limited to certain groups of muscles, e.g., ocular muscles.

The muscles are tender, often swollen, and painful on movement. These symptoms are pronounced in the muscles of respiration, mastication and swallowing. Heavy infestations of the central nervous system causes such symptoms as delirium, somnolence and coma. Myocardial involvement manifested by tachycardia and electrocardiographic changes is common.

3. *Stage of Convalescence.* This starts during the second month as the symptoms slowly subside. Occasionally, rheumatic pains and stiffness will persist. Trichinosis is seldom fatal.

It is unusual to detect all three stages. Many cases remain asymptomatic and are discovered at autopsy.

Pathology. When infestation is heavy, the muscle tissue is pale, soft and granular. Small elongated grayish streaks appear as connective tissue proliferates and the muscle becomes firm. After 6 to 18 months, whitish specks of calcification, which are ovoid, are formed throughout the muscle. Initially, the young trichina lies within the muscle fiber, curls up and evokes an eosinophilic response and granular degeneration with loss of cross striations of the adjacent sarcoplasm. The sarcoplasm undergoes hyaline degeneration, fragmentation, or vacuolization, but the sarcolemmal sheath remains intact, and sarcolemmal nuclei rapidly enlarge and multiply. The sarcoplasm of the intact portion of the fiber becomes basophilic, and the nuclei migrate centrally, congregating about the parasite. Cellular infiltrations consist of both mononuclears and polymorphs, but eosinophils are prominent. When neighboring muscle fibers adjacent to the one harboring the parasite undergo hyaline degeneration and fragmentation, it is considered an effect of toxin elaborated by the parasite. When any part of an invaded fiber is destroyed, [illegible]
to regenera [illegible]
and granularity of the sarcoplasm, hyperplasia of sarcolemmal nuclei, and reforming of cross striations. After the 5th or 6th week, the para-



Fig. 220. Dermatomyositis — microscopic appearance.

sites gradually become encapsulated. Calcification requires from 6 months to 2 years. The larvae may remain viable in the encysted form for many years.

Muscles most susceptible to invasion are, in order of frequency, the diaphragm, extraoculars, tongue, laryngeals, jaw, intercostals, neck, back, abdominals and limbs.

Muscle fiber degeneration and infiltration with inflammatory cells can occur in apparent absence of parasites This suggests that many parasites are destroyed by the inflammatory reaction.

Laboratory Findings Muscle biopsy may reveal the trichinae. A negative finding does not rule out trichinosis, and biopsy should be repeated. Complemental-fixation and intradermal tests, using an antigen prepared from larvae, are of questionable value, because many individuals have become sensitized from previous infection.

Treatment. Prophylaxis demands thorough cooking of pork. Otherwise, symptomatic treatment is given. The prognosis is good

Cysticercosis. This is common in Eastern Europe and India. It is caused by ingestion of eggs of the pork tapeworm, *Taenia solium*. Larval parasites, the *Cysticercus cellulosae*, like trichinosis, invade the intestine and reach the muscles where they cause muscle weakness and tenderness, fever and eosinophilia. Nodules may be palpable in the tongue and other muscles. The encapsulated parasites are situated in the interstitial tissue of the muscle, where they assume a spindle shape and ultimately become calcified. Characteristic shadows of elongated blunt rods are seen on roentgenograms. The diaphragm, the proximal limb muscles and the tongue are preferred sites. Chronic brain lesions causing epilepsy may be the main clinical feature.

INFLAMMATORY MYOSITIS OF UNKNOWN ETIOLOGY

Polymyositis (Dermatomyositis)

This is a rare but serious systemic disorder of collagenous tissues characterized by inflammation and degeneration in skin and skeletal muscle. The cause is unknown. Individuals from 20 to 50 years of age are affected (It is one of a number of conditions classified as diseases of collagen in which the chief characteristics are reaction or hypersensitivity to injury by an unknown agent by fibrinoid de-

generation, cellular proliferation and infiltration, and sclerosis. Other related diseases include disseminated lupus erythematosus, polyarteritis and scleroderma.)

Clinical Picture. Two type can be distinguished: (1) *acute type*, affecting proximal muscles, often associated with skin eruption, and a limited course; (2) *chronic type*, often beginning in periphery of limbs, invariably progressive, dermatitis absent ("polymyositis" is usually applied to the latter type). The clinical picture varies greatly in the acuteness or the insidiousness of onset, the presence or the absence of dermatitis, and intensity of muscular involvement. Although the acute group has a better prognosis with tendency to recovery in a few weeks, respiratory muscle involvement may prove to be fatal. These conditions are characterized by symmetric weakness, tenderness, and induration of large muscle groups, reduction in electric excitability but preservation of faradic response, and liability to fibrous contracture. Visceral lesions include splenomegaly and lymphadenopathy.

1. Acute Type (Dermatitis and Polymyositis). This is commonly seen in children. The acute onset may or may not be associated with fever and leukocytosis. Muscles of the shoulder girdle are often affected first, but soon all the proximal muscles of the extremities become painful, tender, swollen and weak. The overlying skin is reddened and edematous. The face and the neck may display a diffuse erythema and tense edema. Desquamation is common. Other muscles in the extremities are gradually involved. Gradually, the edema and the induration of each muscle slowly subsides, leaving an atrophic muscle, contracted by fibrous tissue. The affected skin may remain indurated and pigmented.

Involvement of mucous membranes causes pharyngitis and stomatitis.

Recovery generally occurs after a long period, but respiratory muscle involvement is of grave import. Residual contracture of muscles causes severe deformity and disability.

2. Chronic Polymyositis. The onset is insidious and affects adults more commonly. Vague prodromal symptoms include fatigability and weakness of certain portions of a limb, usually the lower. Thigh muscles become tender, weak and stiff. Difficulty is experienced in climbing stairs or arising from a chair. The feet and the legs become weak and edematous. Later, similar symptoms are noted in the hands and the forearms. Pain is usually slight or absent. Involvement of the neck and the muscles of deglutition and respiration occurs late. By this time, the peripheral musculature is severely atrophied and contracted (primary generalized myositis fibrosa). Skin lesions are usually absent, but when present they show scaling, induration and pigmentation. Most cases end fatally, usually within 2 years.

Laboratory Findings. Muscle biopsy is diagnostic. The sedimentation rate is elevated. Some cases display an eosinophilia. Hyperglobulinemia and reversal of the A-G ratio is common. Urinary excretion of creatine is increased, but excretion of creatinine is reduced.

Pathology. Early, the muscle is pale red, soft and friable. Later, it is grayish-red or yellowish and firm. Microscopically, widespread degeneration of muscle fibers is striking. Fibers become swollen and rounded in cross section. Myofibrils disappear, leaving a waxy or hyalinized sarcoplasm. This is followed by disintegration by vacuolization, granularity and fragmentation and removal by phagocytes. Diffuse perivascular cellular infiltrations are found. Fibrinoid degeneration of the interstitial tissue and fibrous tissue replacement occur. The sarcoplasm stains darkly basophilic, and its sarcolemmal nuclei are increased in number and lie centrally between the myofibrils or between segmented hyaline masses; this is evidence of an attempt at regeneration. Regenerative signs are uncommon in chronic types.

Cellular infiltration and signs of attempted regeneration are not seen in muscular dystrophy and constitute excellent distinguishing characteristics between the two diseases.

Skin lesions vary. In the more chronic cases, a brownish, tight, shiny, atrophic skin may be indistinguishable from scleroderma.

Prognosis. The disease is usually progressive and fatal in as high as 60 per cent. However, the course is variable and may persist anywhere from weeks to as long as 12 years. As in other collagen diseases, recurrent exacerbations and remissions are typical. Each flare-up may involve a new group of muscles. Occasional cases apparently become stationary. Death occurs by involvement of respiratory

muscles, cardiac muscles, or intercurrent infection.

Treatment. Involved extremities are splinted in the position of function. Light heat, gentle massage and passive exercises through a small range of motion are practiced daily. This program is designed to prevent deformity and contracture in the occasional arrested case. ACTH produces temporary, sometimes dramatic, remissions in some cases.

Interstitial Nodular Polymyositis

Miliary inflammatory nodules develop in the interstitial tissue with destruction of contiguous muscle fibers in skeletal muscles in a group of collagen diseases, including rheumatoid arthritis, rheumatic fever, scleroderma and lupus erythematosus. In any one muscle are seen multiple foci of cellular aggregations consisting chiefly of lymphocytes with other round cells and histiocytes. The infiltrations most frequently are perivascular in location. Muscle fiber degeneration adjacent to the inflammatory focus is evidenced by hyalinization, vacuolization, a loss of striations, and eosinophilic-staining sarcoplasm. Regeneration occurs by formation of muscle giant cells, increase of sarcolemmal nuclei, and formation of basophilic sarcoplasm. Where fibers are extensively destroyed, fibrous tissue replacement occurs.

Clinically, symptoms are minimal or lacking. Perhaps slight and vague muscle aching and tenderness are experienced, and a moderate amount of disuse atrophy is noted.

In rheumatoid arthritis, the muscle fibers are extremely atrophied and thin. In rheumatic fever, the nodules occur in the enveloping muscle sheaths and in tendon bundles near the junction with the fibers rather than in the muscle substance.

Fibromyositis

This is an inflammation of fibrous tissues, particularly those of muscle sheaths, fascia and aponeuroses. The cause is unknown, although cold, dampness and trauma often antedate symptoms. Pain and stiffness about muscles are accentuated by active and passive movement. Firm, tender nodules are often palpable. Pain may be referred to the upper or the lower extremity. The exact histopathology is unknown. (See Fibrositis in section on "The Back").

Polyarteritis

This collagen disease causes fibrinoid degeneration and mononuclear infiltration in small caliber blood vessels. As a result, thrombosis and secondary ischemic necrosis ensue. Muscular involvement by multiple lesions is only one part of generalized disease throughout the body. Grossly, the muscles appear normal. Occasionally, small hemorrhages are visible. Microscopically, multiple well-circumscribed areas of necrosis of muscle fibers are seen. The fibers are swollen, striations disappear, sarcoplasm is eosinophilic and hyaline, sarcolemmal nuclei become pyknotic and smaller. Later, the fibers disintegrate and are removed by macrophages. Replacement takes place by fibrous tissue.

Clinically, the muscles are painful, particularly on motion, tender and weak. Multiple tender spots can be localized, but the periarteritic nodules are microscopic and not palpable.

Epidemic Pleurodynia
(Bornholm Disease, Myalgia Epidemica, Devil's Grip, Epidemic Myalgia)

This is an epidemic acute disease, which is probably of viral origin, and characterized by an acute onset of severe paroxysmal pain about the chest, of fever and frontal headache. It has a seasonal occurrence, almost exclusively in the warm months. Children and young adults are predisposed. The severe pain is invariably at the site of attachment of the diaphragm. It is intensified by sneezing, coughing and deep inspiration. Tenderness is often elicited over the intercostal muscles in the lower thorax. A pleural friction rub may be present. A peculiar characteristic is exacerbation of symptoms at intervals of 1 or 2 days. The condition is benign and subsides in about a week to 10 days. Treatment is symptomatic. An encircling tight bandage about the chest reduces the discomfort. The nature of what appears to be a muscular lesion is unknown.



PLATE 37. Pseudohypertrophic muscular dystrophy (× 85). The muscle fibers are both swollen and atrophic with loss of polyhedral contour. Collagenous and fatty tissue infiltration of endomysium is shown.

MUSCULAR DYSTROPHIES[11, 12]

Progressive muscular dystrophy is a primary degenerative disease of skeletal muscles without evidence of regeneration. In contrast with neural and spinal muscular atrophies, the innervation is intact. This group of diseases is characterized by symmetric distribution of muscular atrophy, preservation of faradic response (proportionate to the residual normal muscle), intact sensation and reflexes, and a heredo-familial incidence.

Etiology is unknown but is generally accepted as hereditary.

Degeneration of muscle due to vitamin E deficiency and to the Coxsackie viruses displays not only degeneration but evidence of regeneration and should not be classified in this group.

CLINICAL FORMS OF MUSCULAR DYSTROPHY

Severe Generalized Familial Muscular Dystrophy (Pseudohypertrophic Muscular Dystrophy of Duchenne). This is a rapidly progressive myopathy, beginning usually in early childhood, with strong familial disposition, occurring predominantly in males, with and without pseudohypertrophy. The disease usually begins before the 6th year. At first the child shows a reluctance to walk or run, he stands and walks unsteadily and falls easily. The muscles usually increase in size but occasionally may decrease. Enlargement of calf muscles, infraspinatus and deltoid muscles is noticeable early. Occasionally, the triceps, the quadriceps and the glutei are affected. Rarely does the initial hypertrophy affect all muscles. The enlarged muscles are firm and resilient but are weaker than muscles of comparable size. The size of muscles increases, but later atrophy sets in. Muscles of the pelvis, the lumbosacral spine and the shoulder girdle are wasted from the onset, causing the characteristic posture and gait. The gait is waddling, the patient standing with the feet spread on a wide base to secure balance. In forward progression, the body inclines from side to side because of gluteus medius weakness. The lumbar spine is extremely lordotic because of weakness of the abdominal wall and the gluteus maximus.

Weakness of hip and knee extensors produces a characteristic movement in arising from a sitting position. The patient pushes his trunk erect by progression, first placing his hands on the legs, then on the knees, and

[11] Erb, W. H: Dystrophia Muscularis Progressiva, Klin and Path Studien. Deutsche Ztschr. Nervenh 1 13, 173, 1891

[12] Adams, R D, Denny-Brown, D, and Pearson, C. M: Diseases of Muscle, New York, Hoeber, 1953.



Fig. 221. Pseudohypertrophic muscular dystrophy. Note the enlarged calf muscles and posterior displacement of the upper trunk assumed for balance.

finally on the thighs. Ultimately, all power in hip, knee, ankle, shoulder and elbow is lost, and atrophy spreads to the periphery of the limbs. Muscles of the hands, the face and the jaw, and also laryngeal, pharyngeal and ocular muscles are relatively spared to the end.

The limbs are flaccid and loose. Shortening and positional contractures appear late. Progression is slow but may be rapid during an intercurrent illness Eventually, the limbs become thin, and the patient is bedridden.

Mild Restricted Muscular Dystrophy (Facio-Scapulo-Humeral Dystrophy of Landouzy and Déjerine[13]). This is less common than the severe generalized type but is not rare. Occasionally, it displays a familial incidence. Both sexes are affected, and the onset occurs at any age but most often between 6 and 20 years. The first complaint is difficulty in raising the arms above the head, often preceded by inability to close the eyes completely because of facial weakness. A "myopathic facies," a sphinxlike appearance with looseness and protrusion of the lips, is characteristic. The patient is unable to purse the lips and to whistle. The eyes cannot be closed against resistance. The lower part of the trapezius and the sternal portion of the pectoral are almost invariably affected, resulting in scapular instability and weakened shoulder abduction. The sternocleidomastoid and all periscapular muscles gradually weaken and atrophy so that the shoulders display exaggerated bony prominences. Biceps and triceps atrophy; the arm appears thinner than the forearm. The disease is often arrested at this stage. Occasionally, slight weakness of the pelvic girdle develops.

The condition is, by comparison, mild and limited and compatible with life. Pseudohypertrophy is rare, but compensatory physiologic hypertrophy may develop in the deltoid and the gluteal musculature.

When muscles of the shoulder girdle and the upper arm are affected without facial weakness, the syndrome is called "juvenile muscular dystrophy of Erb."

Dystrophia Myotonica (Myotonic Dystrophy). This is a progressive, familial myopathy of distal portions of the extremities, the face and the levators of the eyelids. The condition is characterized by markedly delayed relaxation of a muscle after a voluntary contraction. The prolonged contraction may be demonstrated by electrical or mechanical stimulation. This is most pronounced in the hands, the face and the tongue. Gentle movements, such as blinking of the eyes, do not elicit myotonia. A strong voluntary contraction is necessary. The electromyographic characteristics of myotonia are similar to those of Thomsen's disease except that, contrary to the latter, all muscles are not myotonic.

Often the small muscles of the hands and

[illegible] Myopathie [illegible]érédītaire) Altération du Système Nerveux, Comp. rend acad sc. 98 53, 1884.

the forearms are the first to be involved. Muscular wasting usually follows the onset of myotonia by 2 or 3 years. The masseters and the sternocleidomastoids are almost invariably wasted. The peroneals are spared. The disease progresses very slowly. Additional features which typify myotonic dystrophy are endocrine abnormalities, such as testicular atrophy and cataracts. Muscle wasting causes profound general weakness.

Atrophy of the temporal and the masseter muscles produces the characteristic "hatchet face." The lids droop, the cheeks are lax, and the smile is sardonic.

There is no effective treatment.

Progressive Dystrophic Ophthalmoplegia. This condition is a very slowly progressive myopathy which is limited to the levators of the eyelids and the external ocular muscles. It has no orthopaedic significance but is mentioned for completeness.

Laboratory Findings in Muscular Dystrophies

An increased excretion of creatine and decreased creatinine in the urine is characteristic of the dystrophies. Creatine is normally synthesized from glycine and other amino acids, predominantly in the liver, and is deposited in muscle. Creatinine probably arises from muscle creatine by loss of phosphate from creatine phosphate. When muscle is destroyed or wasted, it is less capable of storing creatine, which is excreted instead, rather than being converted to creatinine. It is the decreased excretion of creatinine which is pertinent. On the other hand, increased creatine excretion occurs in a number of diseases which do not



Fig. 222. Myotonia dystrophica. (Case of Dr. Alexander T. Ross)

primarily affect muscles, such as hyperthyroidism.

Normal creatinine excretion is 22 mg. per kg. body weight per 24 hours. Normal creatine excretion is 2 to 3 mg.

The administration of 12 to 18 mg. of α-tocopherol (vitamin E) immediately decreases the creatinuria of healthy children and that which occurs after the administration of glycine. This excretion is similarly inhibited in pseudohypertrophic muscular dystrophy, but weakness and atrophy are unaffected.

Pathology in Muscular Dystrophies

The findings in all 3 types of muscular dystrophy are similar.

Gross Findings. In pseudohypertrophic muscular dystrophy the enlarged gastrocnemii look like fatty tumors, not like muscle Other muscles are small, and their color varies from yellowish to pinkish-gray. The pale translucent appearance, resembling "fish flesh," depends upon the relative amounts of fat and fibrous tissue which replace muscle fibers.

Microscopic Findings. The initial changes consists of swelling and rounding of the muscle fibers, the polygonal contour being lost. Individual fibers become split into daughter fibers, and their interiors become homogenized or hyalinized with loss of striations. Many fibers become smaller than normal. The sarcolemmal nuclei of both swollen and atrophied fibers are increased in number, are larger and assume various shapes. In the myotonic type of dystrophy, the nuclei tend to line up centrally within the fiber. The muscle fibers atrophy and degenerate by vacuolation or granular degeneration, which breaks up the homogeneous appearance. Fat cells accumulate in large amounts between the muscle fibers. *No regeneration is evident anywhere.* (This would be demonstrable by accumulation of sarcoplasm about sarcolemmal nuclei and formation of end buds). Bands of connective tissue and large accumulations of fat cells separate atrophic fibers at a late stage. Motor and sensory nerve fibers are not damaged, and the central nervous system is normal. In the heart, myocardial fibrosis is a common finding.

In the mild restricted type of dystrophy (facioscapulohumeral), an increase in collagenous tissue is a prominent feature, thereby explaining liability to contracture. Deposition of fat occurs but not to the degree observed in pseudohypertrophic muscular dystrophy.

MUSCULAR FIBRODYSTROPHY[14]

Fibrodystrophy of muscle is a chronic nonprogressive condition characterized clinically by generalized weakness and lack of extensibility of all skeletal muscles.

Etiology

The cause is unknown, although a history is often obtained of poliomyelitis or an undiagnosed illness followed by weakness, contractures, and rapid exhaustion on exertion.

Pathology

The muscles display a varying amount of fibrosis and parenchymatous atrophy.

Clinical Picture

The following are typical:

Symptoms. Severe pain and tiredness in muscles after moderate exercise.

Findings. Loss of normal muscle extensibility is evidenced by flattening and limited forward flexion of the lumbar spine, limited ankle dorsiflexion, limited extension at the knee while the hip is flexed, and inability to touch the toes to the floor above the head while lying in the recumbent position. The muscles are soft and lack tone. Tenderness is often found at the musculotendinous junctions. The deep reflexes are diminished. The individual's habitus is asthenic, thin and stooped; *muscle development is poor;* lumbar lordosis is extreme; and the abdomen is prominent. Rarely, an obese or muscle-bound appearance is noted. All laboratory tests are normal.

Treatment

The aim is to lengthen contracted muscles and strengthen their antagonists in an effort to reduce fatigue and improve posture.

[14] Bingham, R : Muscle fibrodystrophy, J. Bone & Joint Surg 29:85, 1947.

CONGENITAL MYOTONIA[15]
(Thomsen's Disease)

Congenital myotonia is a rare, occasionally hereditary, familial disease characterized by delayed muscular relaxation after a strong voluntary contraction. The onset can often be traced back to childhood. A child may be late in learning to stand and walk. Difficulty in movement may be noted as early as 6 years of age, but muscle spasms do not become intense until adolescence or early adulthood. Generalized muscular hypertrophy is usually associated, giving a Herculean appearance. The degree of myotonia varies from case to case and is most prominent in the lower limbs.

Characteristically, after a period of rest, the patient has difficulty initiating movements. The first attempt at movement of the lower limbs causes a painless stiffening contraction which is slow in relaxing. With repeated attempts, successive movements become easier to perform until they occur with ease and are followed by natural relaxation. After resting, voluntary movement again provokes muscular spasm. It is generally necessary to make a strong voluntary contraction to initiate spasm. For example, ordinary blinking occurs naturally, but strong closure of the eyelids will make it impossible to reopen the eyes for a minute or so.

Diagnostic Tests

Percussion myotonia is a spasm with delayed relaxation set up in a muscle by percussing its surface.

Electric stimulation of brief duration initiates a prolonged contraction, although a single shock fails to do so, and repeated electric stimulation abolishes the phenomenon.

Electromyography will reveal, after a strong voluntary contraction, a burst of large action potentials with relaxation followed by small myotonic action potentials. The latter are similar to those in fibrillation.

Thomsen's disease must be differentiated from dystrophia myotonica The latter disease exhibits progression, weakness, arreflexia, hollowing of muscles, especially the masseter, and often a cataract and an atrophied testicle.

The hypertrophied muscles have a greater ability to store creatine and convert it to creatinine. Hence, congenital myotonia has a high tolerance to creatine (in contrast with the muscular dystrophies). No creatine is excreted in the urine; creatinine excretion is high. The serum calcium is normal.

Treatment

Quinine is specific, but it must be continued indefinitely. Calcium also reduces myotonia and may be used as an adjunct to quinine.

FAMILY PERIODIC PARALYSIS[16, 17]

This is a rare, hereditary, familial type of intermittent paralysis which begins in childhood or puberty and recurs for many years. Both sexes are equally affected.

Clinical Picture

Periodic attacks of flaccid paralysis occur, with loss of reflexes and electric excitability affecting the extremities and the trunk. Each attack lasts from a few hours to several days with gradual recovery. It begins with weakness in the back and the thighs and gradually spreads downward to involve the legs and upward to involve the shoulder girdle, the neck and the upper extremities. Muscles supplied by the cranial nerves and muscles of respiration are rarely affected. Premonitary symptoms of excessive perspiration and thirst are common. The attack can be precipitated by excessive cold, carbohydrate ingestion, insulin, ephedrine, and strenuous exercise followed by inactivity. Muscles are normal between attacks. The condition tends to become *benign* with advancing age.

Theory on Pathogenesis

Family periodic paralysis is in some way related to potassium metabolism. Low serum potassium levels occur during an attack, and the administration of potassium is effective in relieving the attack.

[15] Thomasen, E.: Myotonia, Thomsen's Disease, Paramyotonia, Dystrophia Myotonica, p 1, Denmark, Universitetsforlaget i Aarhus, 1948.

[16] Gass, H, Cherkasky, M, and Savitsky, N.: Potassium and periodic paralysis, Medicine 27:105, 1948.

[17] Talbott, J. H: Periodic paralysis: a clinical syndrome, Medicine 20.85, 1941.

Treatment

Small daily doses of potassium can prevent attacks. To relieve the paralysis potassium chloride 10 Gm. is given and repeated in a few hours if necessary. In severe cases, a 2 per cent solution of potassium chloride is given intravenously.

AMYOPLASIA CONGENITA[18]
(Arthrogryposis Multiplex Congenita)

This is a congenital failure of development of skeletal muscles resulting in deforming contractures of joints.

Etiology

This is unknown Hereditary or familial influences are unproved. It has been suggested that the cause is a congenital defect or antenatal degeneration of anterior horn cells plus the maintenance of a fixed position in utero.[19]

Pathology[19]

In the contractured limb, some muscles are normal in appearance, others are small, and still others are absent or replaced by fat and fibrous tissue. The position of contracture deformity depends on one group of muscles overpowering their antagonists, although both groups may be involved. The more affected muscles have a pale pink color. Microscopically, the fibers are small and retain both longitudinal and transverse striations which stain indistinctly. Groups of larger, well-striated fibers may be interspersed among the atrophic ones. The endomysial connective tissue is not increased. Fat cells may be numerous.

In the central nervous system, the anterior horn cells may be reduced in number and size. The brain may be underdeveloped.

Clinical Picture

Involvement may vary from part of one limb to all four extremities. The infant resembles a wooden doll. Fixed deformities occur in any position, but most commonly the arms are rotated internally, the elbows are extended, the forearms pronated, the wrists and the fingers flexed; the hips are flexed and rotated internally; the knees are flexed or extended; and the feet assume a pronounced equinovarus deformity. Clubhand or clubfoot may be present. Erector spinae involvement causes scoliosis. The head muscles are usually spared.

The affected limbs are small in circumference; and the joints, by contrast, appear to be large and fusiform. The joints are not completely ankylosed, some degree of active and passive motion being possible. Muscles are weak, hypotonic, thin and often not palpable. They react poorly to electric stimulation, but the reaction of degeneration is not demonstrable. Tendon reflexes are absent. The joints may be dislocated or subluxated. Other congenital abnormalities may be associated. Intelligence is unimpaired as a rule.

An isolated muscle involvement may be the explanation for various congenital deformities such as clubfoot.

Treatment

Rehabilitation is possible only in mild types with localized involvement. Correction of deformity is attempted by repeated manipulations and application of casts. Tendon transfer procedures to retain the corrected position are rarely effective. Only when a muscle has been proved to be adequate by clinical tests and electromyographic studies can it be used for transfer. Otherwise, one must await bone maturity before attempting arthrodesis.

MYASTHENIA GRAVIS[20]
(Erb-Goldflam's Disease[21, 22])

Myasthenia gravis is a disease characterized only by muscle weakness. In its mild form it is extremely common. Frequent involvement of craniopharyngeal and intercostal muscles, or weakness of the back and the lower extremities, causes confusion with poliomyelitis and muscular dystrophy.

Etiology

Females are predisposed in the ratio of 2 to 1. The condition occurs at any age, but individuals in the fourth decade are favored.

18 Middleton, D S: Studies on prenatal lesions of striated muscle as a cause of congenital deformity, Edinburgh M. J 41:401, 1934

19 Adams, R D, Denny-Brown, D., and Pearson, C. M: Diseases of Muscles, New York, Hoeber, 1953.

20 Tether, J E: Orthopaedic Aspects of Myasthenia Gravis, Am. Acad Orthop. Surgeons, Lect. 9 171, 1952

21 Erb, W Arch. Psychiat 9 336, 1878

22 Goldflam, S · Neurol. Ztschr 21:97, 1902.

Heredity plays no part. A myasthenic mother may give birth to a myasthenic infant. The fact that myasthenia in the infant is of short duration suggests that some causative circulating substance crosses the placental barrier. Infection and injury may precipitate or aggravate the disease. Menstruation usually intensifies symptoms; pregnancy may effect a remission. Because psychic stress often greatly aggravates the muscle weakness, and because thymicolymphatic involvement is seen in certain severe cases, an endocrine etiology in which an adrenal stress mechanism operates is a possibility.

Physiology

Normally, a motor-nerve impulse liberates acetylcholine at the myoneural junction. Acetylcholine effects a muscular contraction, then is destroyed by cholinesterase. Therefore, myasthenia gravis may be due to (1) insufficient synthesis of acetylcholine, (2) excess cholinesterase, or (3) a curarelike blocking agent which decreases the receptiveness of muscle to acetylcholine.

The theory of a blocking agent is supported by the following facts: (1) the myasthenic resembles the normal curarized individual; (2) myasthenics are hypersensitive to curare, from 10 to 50 times normal; (3) if in a severe myasthenic an extremity under a tourniquet is exercised and then the tourniquet removed, general fatigue will be aggravated; (4) normal muscle may release a curarelike agent in extreme fatigue;[23] (5) electromyogram of curarized normal muscle is identical with that of a myasthenic.[24]

The thymus is often found to be enlarged, rarely neoplastic The suggestion that it produces a curarelike agent is unfounded.

Pathology

The muscles appear normal grossly except for some atrophy of disuse. Microscopically, only small collections of lymphocytes, termed "lymphorrhages," are occasionally found in the interstitial tissues.

[23] Torda, C Release of curare-like agent from healthy muscle and its bearing on myasthenia gravis, Proc Soc Exper. Biol & Med 58 242, 1945.

[24] Harvey, A M, and Masland, R. L The electromyogram in myasthenia gravis, Bull. Johns Hopkins Hosp 69 1, 1941.

Clinical Picture

Abnormal fatigue of voluntary muscles is the one main symptom. It is aggravated by exertion, relieved by rest, worse at the end of the day, and involves a single muscle, group of muscles, or the entire musculature.

The extra-ocular and lid muscles are the first to be involved in half the cases, causing ptosis, strabismus and conjunctivitis from incomplete lid closure.

Facial muscle weakness is frequent. It causes a characteristic sad expression.

Fatigue of jaw muscles is noted when masticating tough meats.

Tongue muscle fatigue causes dysarthria. The patient complains that his tongue feels thick, and he talks as though he has his mouth full of hot potatoes.

Dysphagia caused by pharyngeal muscle weakness is worse toward the evening meal. Fluids often regurgitate through the nose or are aspirated. Pneumonic infection is a constant danger.

Neck muscle fatigue may be prominent. The patient is unable to hold his head erect.

Fatigue of the upper extremities is at first noted when attempting upward reaching movements. Women complain of difficulty in caring for their hair. Men may experience tiredness when shaving.

A sense of heaviness in the chest or a feeling of insufficient deep breathing are complaints due to fatigue of respiratory muscles. When these symptoms become severe, respirator care is necessary.

Involvement of the lower extremities is first noted in the complaint of inability to climb stairs, or a sense of collapsing of the knees after a short period of walking.

The spinal and the abdominal muscles are also involved.

Diagnostic Tests

Neostigmine bromide, a parasympathetic stimulant, is given in doses of 15 mg. 3 times a day. It is important that it be administered after meals, because its absorption from the fasting empty stomach is too rapid. Side-effects of abdominal cramping, diarrhea, or nausea are noted only in mild cases or normal individuals and are counteracted by belladonna or atropine

If the patient is myasthenic, neostigmine will effect an increase in strength of all affected muscles. It may be necessary to increase the dosage gradually before this effect is noted. If, on the other hand, only side-effects appear the patient is not a myasthenic.

The Myasthenic Reaction of Jolly. When a myasthenic muscle is stimulated by faradic current, the first few contractions will be strong, then subsequent ones show gradually lessened response.

Provocative tests with curare or quinine must be used when a very mild case is suspected or when long-standing cases render the muscles incapable of response to neostigmine. It is important to remember that myasthenics are extremely sensitive to curare. One tenth or less of the minimal curarizing dose for a normal person of the patient's weight is injected slowly intravenously. A transient aggravation of myasthenic symptoms and findings occurs (ptosis of eyelids may be evident) The curare is overcome by intravenous injection of neostigmine 0.5 mg.

Clinical Course

The onset may be gradual or sudden. Remissions usually last weeks, months, or years; in most cases the disease is not progressive. Most patients require less neostigmine as time goes on. An occasional case becomes neostigmine-resistant, weakness worsens, and respiratory failure may occur. Use of a respirator may carry the patient past the crisis.

Treatment

Neostigmine bromide in syrup base containing 15 mg. to the dose is given 3 times a day after meals. Absorption from the gastrointestinal tract and requirements of the individual are variable. Only a sufficient amount to arrest symptoms is necessary. Overdosage may cause muscular twitching and weakness due to excess acetylcholine. It may be preferable to allow the patient to judge the need for medication, e.g., when symptoms of slight heaviness in the eyelids, the face, or the arms appear. Ephedrine in a dose of 24 mg. 3 times a day is a useful adjunct to neostigmine therapy.

Various orthopaedic problems present themselves. Muscular dystrophy and poliomyelitis are common diagnoses. Weakness of back muscles may cause a severe lumbar lordosis (to sustain balance) and complaints of pain in the back and the lower limbs, always worse toward the end of the day. Oral neostigmine is diagnostic and therapeutic.

A peculiar waddle suggestive of hip instability is caused by weakness of the pelvitrochanteric group of muscles. The Trendelenburg may not be noticeable except after a period of exercise.

Any complaint of weakness and fatigue, whether of the neck, the back, or the extremities, or inexplicable falling episodes should receive the benefit of a therapeutic test. If myasthenia gravis is diagnosed, treatment should consist of neostigmine and encouraging activity. Brace supports should be avoided, as they promote muscle atrophy.

Removal of an enlarged thymus has been tried with questionable success and a high mortality. In view of the relatively good prognosis with drug therapy, the operation is not indicated.

16

Fibrous Diseases

FIBROUS DYSPLASIA OF BONE[1,2]

Fibrous dysplasia of bone is a relatively rare condition characterized by fibrous tissue replacement of the skeleton. It may be monostotic (confined to one bone) or polyostotic (situated in many bones).

[1] Albright, F., Butler, A. M., Hampton, A. O., and Smith, D.: Syndrome characterized by osteitis fibrosa disseminata, areas of pigmentation, and endocrine dysfunction with precocious puberty in females, New England J. Med. 216:727, 1937.

[2] Pritchard, J. E.: Fibrous dysplasia of bone, Am. J. M. Sc. 222:313, 1951.

Etiology

The cause is unknown. A plausible explanation is the hyperostosis which develops at the base of the skull where it can compress the pituitary and the hypothalamus. The condition begins in childhood, and its progress is halted when growth has been completed. Both sexes are equally affected.

Pathology

The lesion may implicate one, several or many bones and often favors one side of the body. Bones of the extremities and the base



Fig. 223. Fibrous dysplasia of bone and a pathologic fracture.



Fig. 224. Fibrous dysplasia of bone.

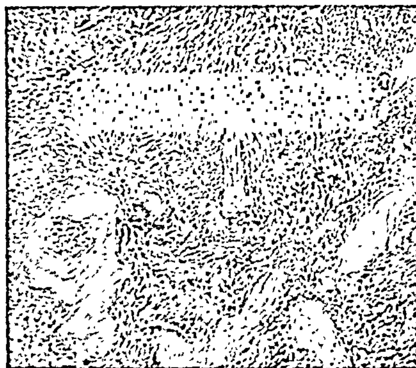


PLATE 38. Fibrous dysplasia. (× 120)

of the skull are predominant locations. The epiphyses are uninvolved. Basically, the bony structure is replaced to a variable degree by avascular fibrous tissue.

Gross. The bone is irregular and often bent. The cortex is thin and bulged outward by the underlying abnormal tissue. Removal of the cortex exposes a white or brown tough fibrous tissue which cuts with a gritty resistance. The affected bone is usually shortened but occasionally may be longer than normal. Pathologic fractures occur but heal readily with deformity. Outward bowing of the shaft and a varus deformity of the neck produces a characteristic "shepherd's crook" deformity of the femur. A hyperostosis often develops at the base of the skull and may obliterate the sinuses and encroach upon the foramina.

Microscopic. Dense bundles of collagenous tissue form whorls and enclose thin bony trabeculae. Osteoclastic activity is minimal (as contrasted with pronounced osteoclasis in hyperparathyroidism). Occasional islands of cartilage are seen. Cyst formation is infrequent. Rarely, lipoid-laden histocytes are present.

Symptoms. The dysplasia starts in early childhood but is usually mild and asymptomatic, being discovered on x-ray survey. When the condition is severe, a constant, dull, aching pain is complained of. A bending *deformity* of an extremity may gradually develop or follow a fracture. *Pathologic fractures* are frequent but unite readily. The involved extremity may be longer or shorter than on the opposite side. In the skull, a hyperostosis frequently develops at the base and the maxilla and causes *asymmetry of the head and the face.* Characteristic large, brown, irregular patches of skin *pigmentation* are usually associated with the polyostotic type.

Sexual precocity is typical in females, evidenced by early menstruation, breast development and epiphyseal closure. The condition is often *unilateral* in distribution. Activity is halted after cessation of growth.

Albright's syndrome consists of the combination of unilateral polyostotic fibrous dysplasia, pigmentation, and sexual precocity occurring in a female.

LABORATORY FINDINGS

The serum calcium, phosphorus and alkaline phosphatase are normal. In severe cases, the alkaline phosphatase may be elevated.

ROENTGENOLOGIC FINDINGS

The typical lesion is a well-circumscribed area of homogenous rarefaction occupying a portion of the shaft of a long bone. The "ground glass" lesion is in direct contrast with the denser well-trabeculated adjacent normal

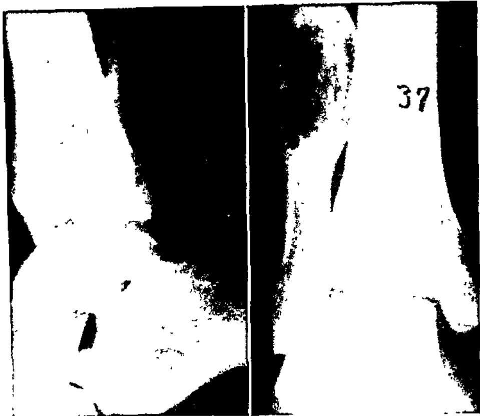


Fig. 225. Fibrous "cyst" of fibula.

bone. The overlying cortex is thinned and expanded. A cyst is suggested by the localized radiolucent area. Pathologic fractures cause shortening, bowing and distortion throughout the lesion. This may result in a loculated, cystic appearance.

The skull is characteristic. A dense hyperostotic formation develops and enlarges at the base, obliterating the sinuses and thickening the diploe.

Differential Diagnosis

The main conditions to be differentiated are hyperparathyroidism, osteogenesis imperfecta and neurofibromatosis.



Fig. 226. Monostotic form of fibrous dysplasia. Clinically, only a local swelling with mild discomfort. Progress slow. [illegible]

[illegible] medullary, expansile, erodes, expands, and thins cortex. Grossly, grayish or yellowish fibrous tissue was found and was gritty on cutting. The lesion was cleaned out and replaced with a large cortical and multiple cancellous grafts. (N. U. Case No. 220)



Fig. 227. (*Left*). Monostotic fibrous dysplasia. Extensive involvement jeopardized the integrity of the shaft, necessitating resection of the lesion and replacement with bone chips and a long tibial cortical graft.



Fig. 228 (*Right*). Nonossifying fibroma of bone.



Hyperparathyroidism. The serum calcium is high, the serum phosphorus is low, and the alkaline phosphatase is high. The condition is more generalized and cystic, and the lamina dura of the teeth is absent. Fibrous dysplasia has normal serum findings (except an elevated phosphatase in severe cases), an earlier onset, osteoporosis is not generalized, the base rather than the vault of the skull is affected, pigmentation is present, and various endocrine disturbances are common.

Osteogenesis Imperfecta. Multiple fractures, blue sclerae and deafness are characteristic. The bones are slender in contrast with widened shafts of fibrous dysplasia. The osteoporosis is generalized. Vertebral bodies are compressed. In fibrous dysplasia, the spine is rarely affected.

Neurofibromatosis. The lesions of bone predominantly involve the lower end of the femur

Fig. 229 (*Left*). Nonossifying fibroma.

and the upper end of the tibia; there is no tendency to unilateral distribution; the pigmented skin patches are smoother in outline; and multiple nodules in the subcutaneous tissue are often palpable. Microscopically, the tissue when stained by the method of Rio Hortega displays specific cells, the lemmocytes.[3]

Treatment

A weight-bearing extremity may require brace protection. Fractures often are followed by deformity and should be treated by open reduction and bone graft fixation. A large lesion which jeopardizes the integrity of the shaft requires curetting and obliteration with bone chips. When the limb is severely involved, deformed and shortened, amputation and fitting with a prosthesis may seem to be desirable.

Sarcomatous degeneration has rarely been reported. Persistent pain and suggestive rapid changes on x-ray examination should be pursued further by biopsy.

NONOSTEOGENIC FIBROMA OF BONE[4,5]

Nonosteogenic fibroma of bone is a benign, well-circumscribed, fibrous growth within a small area of a long bone. It is probably a localized form of fibrous dysplasia. It is seen in older children and adolescents, usually between the ages of 8 and 16. Both sexes are equally affected. The lesion occurs in the metaphysis of a long bone, most often in the lower limb, and predominantly at the lower end of the femur and at both ends of the tibia and the fibula. Frequently, it is symptomless and is discovered accidentally; or pain over a palpable, tender, bony swelling is the original complaint; or a pathologic fracture initiates symptoms.

[3] Valls, J., Polak, M., and Schajowicz, F.: Fibrous dysplasia of bone, J. Bone & Joint Surg. 32A:311, 1950.

[4] Jaffe, H. L., and Lichtenstein, L.: Non-osteogenic fibroma of bone, Am. J. Path. 18:205, 1942.

[5] Hatcher, C. H.: The pathogenesis of localized fibrous lesions in the metaphyses of long bones, Ann. Surg. 122:1016, 1945.

Roentgenologic Findings

These are distinctive. The lesion is sharply defined, translucent, loculated, and possesses a thin border of increased density. It is oval typically, about 1 to 1½ inches in length, and its long axis lies in the long axis of the bone. The location is eccentric, the overlying cortex being thin and expanded. In slender bone, the fibroma occupies the entire width of the shaft.

Pathology

Grossly, a thin cortex encloses a soft or a tough, rubbery, gray-yellow or reddish-brown tissue.

Microscopically, fibrous tissue of varying cellularity is seen. A more cellular fibrous tissue contains plump spindle cells, is quite vascular, and deposits of hemosiderin produce the reddish-brown color. A few giant cells may be seen. Trabeculae are conspicuously absent. When a tissue is more fibrous and in whorls, it is less cellular and less vascular.

History of the Lesion

The tendency is for the earliest lesion to appear near the epiphyseal plate, enlarge and move away from the plate, then gradually become smaller, indistinct and then disappear. The lesion is rarely seen in late adult life. Opinion is divided on the method of obliteration: (1) metaplasia to bone and (2) ingrowth from surrounding bone.

Theories of Etiology

Jaffe and Lichtenstein feel that this is a benign tumor. However, spontaneous disappearance of the lesion seems to refute this theory. Hatcher suggests that this "fibrous metaphyseal defect" might be a local disturbance of bone growth originating at the epiphyseal plate.

Treatment

The indications for treatment are (1) persistent pain and tenderness, and (2) extensive involvement which might jeopardize the integrity of the bone. Treatment consists of complete excision and curetting the lesion down to normal bone and filling the defect with bone chips. Otherwise, the lesion is considered as benign and, given sufficient time, should disappear.

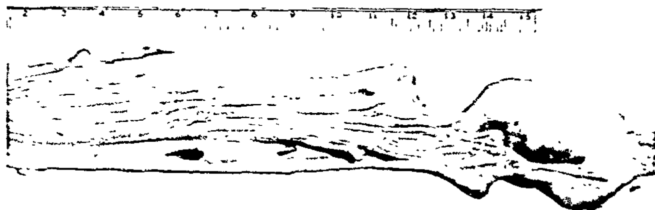


FIG. 230 Neurofibromatosis. Multiple involvement of the nerve roots at the cauda equina. Patient died at the age of 42 from sarcomatous degeneration of a neurofibroma of the brain. (Pathology Department, Mount Sinai Hospital, Chicago)

CONGENITAL NEUROFIBROMATOSIS[6, 7]
(von Recklinghausen's Neurofibromatosis)

This congenital hereditary condition which can appear at birth or at any time thereafter is characterized by the development of one or many neurofibromata, skin lesions, enlargement of a limb, and secondary skeletal changes.

CHARACTERISTICS

1. **Strong Hereditary Tendency.** It has been found in as many as 6 generations.

2. **Skin Lesions**

A. *Pigmented.* Café-au-lait spots

B. *Cutaneous Fibromata.* Flat or raised, soft, multiple. Appears as verrucae or fibroma molluscum.

3. **Multiple Neurofibromata.** Consists of connective tissue derived from endoneurium or perineurium which form whorls of fibrous tissue interspersed with sparse nerve fibers. These can occur wherever a peripheral nerve exists, the symptoms produced depending on the site.

A. *Subcutaneous.* These are tender, painful, palpable nodules varying from pea-sized to a very large tumor occupying almost the entire part of the extremity. Rarely, a plexiform type forms, consisting of tortuous cordlike tumor masses representing enlargement of every nerve filament of an extremity.

B. *Subperiosteal.* The tumor by compression causes local bone resorption, then excites the periosteum to produce new bone which envelops the tumor, and a subperiosteal bone cyst results.

C. *Endosteal.* No typical lesion within the bone has ever been described. However, involvement is suggested by the frequently associated bone changes. The bone becomes porous and plastic and increases in length. Rarely, growth is retarded, supposedly by tumor interference at the epiphyseal plate. Bone cysts and pseudarthrosis of the tibia have been reported.

D. *Intraspinal.* An hourglass-shaped tumor of the nerve root may erode and enlarge the intervertebral foramen. Within the spinal canal, the interpedicle space is widened, and the posterior surface of vertebral bodies displays smooth indentations of erosion. The cranial nerves may be involved.

4. **Skeletal Changes.** These are caused by external pressure and erosion, direct intraosseous involvement, and irritation or damage to epiphyseal longitudinal growth.

A. *Long Bones.* Increased rate of growth in length is characteristic. A smooth erosion of the cortex is apparent at the tumor site. A cortical cyst may develop. The general bony structure is usually porotic. Rarely, increased density and multiple bony cysts are found.

B. *Spine.* A sharply defined scoliosis fre-

[6] Fairbank, H. A.: J. Bone & Joint Surg 32B 266, 1950.

[7] McCarroll, H. R. Clinical manifestations of congenital neurofibromatosis, J. Bone & Joint Surg. 32A:601, 1950.

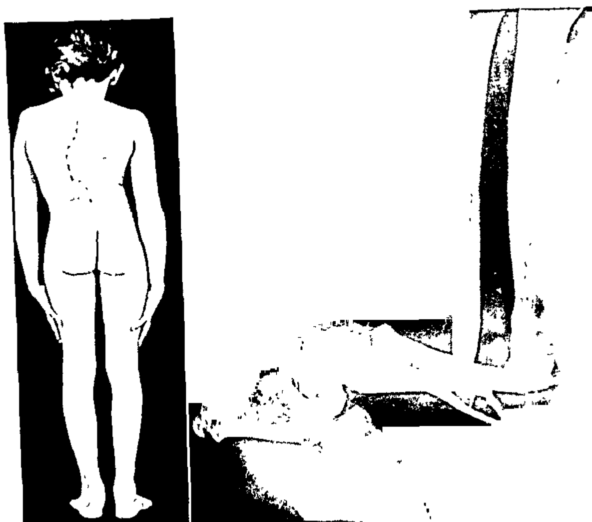


FIG. 231. Congenital neurofibromatosis. (*Left*) Extreme congenital scoliosis in a child. The angulation is acute and usually rapidly progressive and disabling. Fusion should be done promptly. (*Right*) Pseudarthrosis of tibia at site of a cystic lesion. (McCarroll, H. R.: J. Bone & Joint Surg. 32A:601)

quently develops, usually at the lower dorsal area. A kyphosis of the affected segment is associated. The kyphoscoliosis is progressive, may be crippling and demands surgical treatment. The enlargement of the intervertebral foramen, widening of the interpedicle space, and erosion of the posterior surface of the vertebral body has been mentioned.

5. **Elephantiasis.** Enlargement of a part or all of an extremity is caused by a combination of factors, including diffuse hypertrophy of all soft tissues, edema due to lymphatic involvement, hemangioma formation, a large neurofibroma, and enlargement of the bone.

PATHOLOGY

Grossly, the neurofibromatous nodule is a firm, dense fibrous appearing structure, usually nonadherent to surrounding structures.

Microscopically, the tissue is almost identical with that of a fibroma. The typical dense strands and whorls of fibrous tissue are seen everywhere and may be easily confused with the picture of fibrous dysplasia when the specimen has been removed from bone. However, in von Recklinghausen's disease, when the tissue has been stained by the method of Rio Hortega, specific cells, the *lemmocytes,* are identified [8] These are cells of neural (ectodermal) origin, are elongated, have rodlike nuclei and scarce cytoplasm which terminates

[8] Valls, J, Polak, M., and Schajowicz, F.: Fibrous dysplasia of bone, J. Bone & Joint Surg 32A·311, 1950.

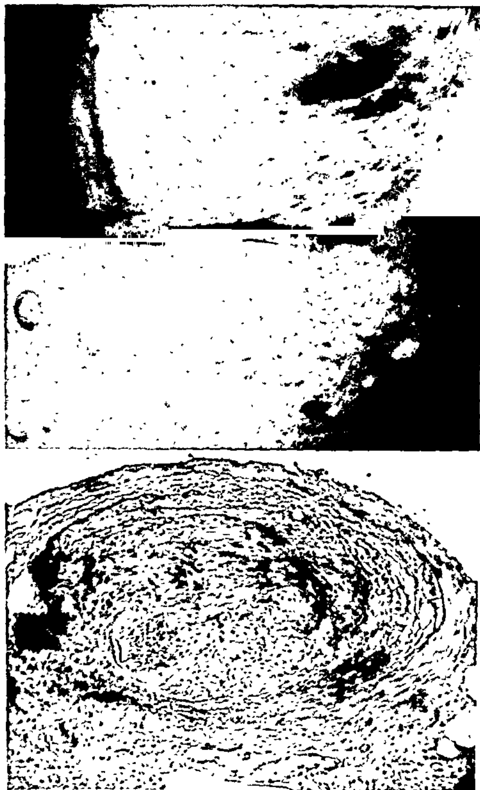


FIG. 232. Congenital neurofibromatosis. (*Top*) Typical multiple subcutaneous tumor nodules. These are composed of nerve tissue covered by areolar tissue and skin. (*Bottom*) Low-power magnification of enlarged nerve fiber removed from a plexiform neurofibromatous mass. (McCarroll, H. R.: J. Bone & Joint Surg. 32A:601)

in 2 or 3 prolongations, designated by Rio Hortega as "neuritides" The mesodermal cells also found are fibroblasts and histiocytes.

Clinical Picture

The asymmetry of the face, the body and the limbs is apparent. Symptoms of pain and disturbance of function are related to the particular peripheral nerve involved. The lesions may or may not be progressive when seen. However, the exception is the kyphoscoliosis which increases and is painful and disabling.

Treatment

Complete excision is the only treatment. The elephantiasis is treated by repeated re-



Fig. 233. Congenital neurofibromatosis. (*Top*) Diffuse soft tissue hypertrophy and increased length of the lower extremity. Picture at the right reveals typical tumor mass of hypertrophied soft tissue exposed at operation. It is not encapsulated and is superficial to the deep fascia. (*Bottom*) Hypertrophy of left thumb and index finger and corresponding portion of the hand. Massive skeletal hypertrophy in the involved segments. (McCarroll, H. R.: J. Bone & Joint Surg. 32A:601)

section of hypertrophied soft tissue as well as the tumors Painful tumors within the spine are removed after adequate decompression Scoliosis demands surgical correction because conservatism will not halt its progression. Surgery in these patients is fraught with the dangers of hemorrhage and shock to which they are particularly susceptible.

17

Unclassified Diseases of Bone

PAGET'S DISEASE
(Osteitis Deformans)

Paget's disease is a very common chronic affection of the skeleton occurring in individuals past middle age. It is characterized by the development of thickening and deformity of osseous structures and by complications of fractures and malignant degeneration.

Etiology

The actual cause is unknown. Endocrine and metabolic disturbances are ruled out, because even in the presence of extensive involvement many bones are free of disease. Males are affected more often than females. Occasionally, a hereditary influence is noted. Most cases are observed after 50 years of age. Schmorl believed that 3 per cent of everyone over 40 has osteitis deformans.

Pathology

Early, the basic process is one of extensive osteoclastic destruction accompanied by increased vascularity and fibrosis. Trabeculae are thinned, and haversian canals are enlarged. Periosteal new bone and, to a lesser extent, endosteal new bone forms and thickens the cortex. This fibrous bone is soft and yields easily to stresses and strains of weight-bearing. Next, the stage of repair begins. Large numbers of osteoblasts are seen laying down thick, coarse trabeculae of new bone, replacing the old cortical trabeculae and the recently formed trabeculae beneath the periosteum. Repair and destruction are taking place at the same time As new bone is being formed in some places, it is destroyed in others. The process is disorderly and disorganized. The new trabeculae are distorted in shape and direction and often are laid down about trabeculae which are old and incompletely absorbed. The trabeculae are joined by deeply staining cement lines arranged in bizarre fashion, referred to as the characteristic mosaic pattern of the disease



Plate 39. Paget's disease (× 85) Showing the mosaic structure of thick trabeculae, very active bone formation, and vascular fibrous structure of the medullary spaces.

Fig. 234. Paget's disease, affecting mainly the spine, the pelvis and the left femur.



There appears to be no effort at formation of haversian systems. In contrast with the soft fibrous bone, dense mosaic bone is brittle and easily fractured. Bone repair is stimulated by activity, so that those bones which are subjected to pressures of weight-bearing and stresses of muscular activity display early and intense new bone formation. Thus, the long bones of the lower extremities and the vertebrae exhibit the dense bone of an active reparative process which keeps pace with the destructive process. In the skull, where stresses and strains are at a minimum, repair lags considerably behind in the early stages, and destruction predominates as revealed by a sharply demarcated soft osteolytic vascular area. The outer skull surface becomes greatly thickened by periosteal fibrous bone. Replacement by mosaic bone is delayed but inevitably occurs.

Periods of remission are thought to occur. At this time the microscopic appearance is one of thickened, irregularly disposed, mosaic bone, acellularity, and marrow spaces occupied by avascular fibrous tissue.

Although any bone may be affected, those involved in the order of greatest frequency are the pelvis, the femur, the skull, the tibia and the spine. The bones of the hand and the foot are seldom affected (a differential point from polyostotic fibrous dysplasia). The condition may be confined at first to a portion of one bone (usually the tibia or the femur) and spreads gradually to the rest of the bone. At its advancing sharply delimited edge may be observed microscopic evidence of early osteoclastic bone resorption. It may remain within that bone for years as a solitary lesion, but, almost invariably, other bones eventually become affected. Although the condition becomes widespread, many areas escape, and the distribution is asymmetric.

Grossly, the bones are thickened and display irregular rough surfaces The long bones become bowed anterolaterally in response to pressure of weight-bearing. Over the convex outer cortex are often seen transverse fissures constituting incomplete fractures which have healed with a minimum of callus. Frequent refracturing and consequent outward bowing cause actual lengthening of the shaft. (A complete transverse fracture may occur and heal.) The cortex is thickened not only externally but also internally with encroachment upon the medullary cavity. The spongiosa is composed of coarse strands of bone.

The bones of the vault of the skull are affected. Fortunately, only the external cortex is thickened. However, the distinction between internal and external tables is lost, and the sutures become obliterated. The bones of the face are rarely affected.

Epiphyses often are involved with the result that weight-bearing articular surfaces deteriorate rapidly, and typical osteoarthritic joints are produced.

Clinical Picture

Very commonly the disease exists asymptomatically in one or several bones and is discovered accidentally on x-ray examination.

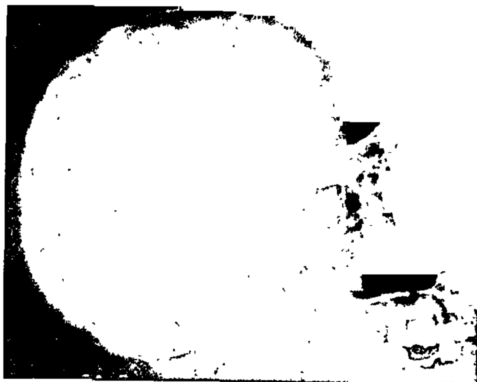


Fig. 235. Paget's disease of the skull. Note the wooly or cottonball spots and loss of distinction between the inner and the outer tables. Large well-demarcated areas of osteoporosis circumscripta are observed in the frontal and the occipital regions.



The tibia or a vertebra are favorite sites for the early monostotic form of the disease.

The onset is very insidious. The patient is of middle or advanced age, and more often a male. The initial complaint may be pain in the area of bone involvement. This pain varies in type and intensity. It may be a dull ache, a "neuralgic" discomfort, or a sharp stabbing sensation. It may be intermittent or constant and aggravated by various causes. Spontaneous subsidence of the discomfort is not unusual. When pain is felt over the tibia, perhaps the bone is found to be enlarged and bowed. The overlying skin is often warm to the touch, but the bone is not tender. Headache is a common symptom. The head gradually enlarges, necessitating frequent changes of hat size. Backache is a common accompaniment of involvement of the spine.

Gradually, over a number of years the involvement often becomes more widespread. The enlarging head is a sharp contrast with the relatively small face. One or both legs and/or thighs become bowed anterolaterally, the enlarged tibiae increasing the circumfer-

Fig. 236. Paget's disease of the femur. The changes at the upper end of the tibia are reactive degenerative changes in response to irregularity of the lower surface of the femur.



Fig. 237. Paget's disease of the pelvis.



Fig. 238. Osteogenic sarcoma arising in Paget's disease of the tibia. Sun-ray appearance is evident.

ence of the legs. A diffuse kyphosis develops throughout the spine. The chest is narrowed in its transverse diameter. A characteristic picture is produced in the advanced stage. The patient stands in a crouched position with the enlarged head thrust forward, the trunk bent, and the lower extremities widely bowed outward. Gait is slow. Muscle weakness and fatigue are evident.

Although the cranium is thickened, compression of the brain and narrowing of foramina never occur. Deafness is frequent but is due to otosclerosis. Visual impairment is caused by arteriosclerosis.

Although seemingly disabled, the victim of Paget's disease may live out his normal life's expectancy unless he develops sarcomatous degeneration.

Complications[1, 2]

The commonest complication is *fracture*, especially of weight-bearing long bones. Fracture is thought to be more common in the vascular, soft stage. At this time multiple incomplete transverse fissures are found over the apex of the convexity of the bowed shaft These heal rapidly. Occasionally, however, a complete transverse fracture can occur and heals readily, although with a minimum of



Fig. 239. Paget's disease with malignant degeneration.

[1] Jaffe, H. L · Paget's disease of bone, Arch Path 15 83, 1933.

[2] Lake, M.: Studies of Paget's disease, J. Bone & Joint Surg. 33B:323, 1951.

callus. When fracture occurs in the late stage of dense bone, union is delayed.

At least 5 per cent of cases undergo *malignant degeneration*. The tumor is osteogenic sarcoma, fibrosarcoma, or round cell sarcoma. Its development is often heralded by an increase in pain and rapid increase in size of a bone. It is often multicentric. Osteolytic changes are noted in roentgenograms. The tumor is highly malignant. It is said to arise more commonly in the vascular phase, and its origin is frequently subperiosteal. Biopsy confirms the diagnosis.

Roentgenologic Findings[3]

The roentgenologic appearance varies, depending upon the relative amount of destruction and reconstruction. Actually, even when the disease is detected early in a long bone some bony formation is already taking place, especially by periosteal new bone formation thickening of the cortex Only at the advancing sharp edge of the lesion can one see the rarefaction, the earliest stage of the lesion. Eventually, the cortex is thickened, the medulla often narrowed, the shaft bowed, and thick coarse strands of bone develop which vary from a honeycombed or spongy appearance to large dense strands of bone. Small transverse radiolucent lines often interrupt the cortex over the convexity, representing incomplete fractures or Looser's zones of transformation.

In the pelvis, trabeculation is very coarse and thick and most conspicuous at the periphery as multiple, parallel, curvilinear lines below the iliac crest. An early finding is exaggerated trabeculae above the acetabulum. Occasionally, the transverse diameter of the pelvis is narrowed by pressure of weight-bearing during the "soft period." The pubic and the ischial bones are widened. This finding is especially important when the bone is quite dense and must be differentiated from osteoplastic metastases.

In the spine, one or more vertebrae are affected. Coarse trabeculae form parallel with the periphery and densely outline the vertebral body. The remainder of the body in the early stages appears decalcified and soft and is often compressed and widened in its transverse diameter. However, encroachment on the spinal canal practically never occurs. Ultimately, the body becomes quite dense and devoid of the trabeculated architecture. Its contour often is square.

The best example of the destructive phase is seen in the skull. Early, a well-delineated decalcified lesion is seen affecting chiefly the outer table. This is known as *osteoporosis circumscripta*. A thin layer of periosteal bone may overlie the lesion. Eventually, multiple spots of increased bone density develop within the rarefied area. These new bone densities often are round with ill-defined edges and are described as "cotton-ball" spots. Ultimately, the densities extend irregularly throughout the skull. The outer table thickens, and the distinction between outer and inner tables is lost.

When a sarcoma develops, often it is revealed by a saucer-shaped erosion of the outer cortex, usually signifying a fibrosarcoma; or the usual signs of osteogenic sarcoma, including irregular areas of destruction within the shaft, periosteal new bone, sun-ray appearance, etc.

Laboratory Findings[4]

The serum calcium and phosphorus are usually normal. The alkaline phosphatase is higher in Paget's disease than in any other condition. Its level is an index of bone formation. Conversely, the disease may be regarded as in a stage of remission when the serum alkaline phosphatase level is normal.

The Effect of Immobilization on Paget's Disease

When a bone affected by Paget's disease is immobilized, the stimulus for bone formation ceases, but bone destruction continues. Therefore, the serum alkaline phosphatase level is decreased, and the urinary excretion of calcium is increased. If the quantity of calcium exceeds that which the kidney is capable of eliminating, a hypercalcemia results. Osteoporosis is extreme. The symptoms of hypercalcemia include nausea and vomiting, dryness of the nose and the throat, and difficulty in swallowing.

[3] Pugh, D G: Roentgenologic Diagnosis of Disease of Bones, Baltimore, Williams & Wilkins, 1951

[4] Albright, F., and Reifenstein, E C, Jr.: The Parathyroid Glands and Metabolic Bone Disease, Baltimore, Williams & Wilkins, 1948.

When a fracture has occurred, similar to any injury, the 17-ketosteroid excretion is depressed. This altered adrenocortical function curtails tissue formation, thus further encouraging osteoporosis. However, the site of injury seems to be unaffected, and fractures heal.

The danger of "chemical death" exists. Immobilization of a patient with Paget's disease, who already may be ingesting large quantities of milk, may encourage a hypercalcemia, renal calculi, metastatic calcification in the lungs, anuria, and death when the patient is bedridden or his activity is curtailed. Hypercalcemia and hypercalciuria must be controlled by a low calcium diet, magnesium salts and fluids.

Treatment

Since the original defect is bone resorption, it may seem appropriate to furnish adequate calcium and phosphorus, dilute acid and bile salts to aid calcium absorption, and vitamin D to promote bone formation. Sex hormones, especially estrogen in the female, benefit bone repair. Of course, the patient must be kept constantly active. This is particularly true after a fracture. If possible, the use of an intramedullary nail will permit full use of the extremity. Severe deformity of the tibia or the fibula may be corrected surgically, preferably by the delayed osteotomy method of Moore

Decalcifying Agents. Stein[5] has used compounds of magnesium during the osteoblastic phase. Magnesium acts as a decalcifying agent by increasing both fecal and urinary excretion of calcium and phosphorus. The patients seem to gain in strength and endurance, the serum alkaline phosphatase declines, and the urine Sulkowitch test shows large amounts of calcium being excreted He recommends the following program:

Osteolytic Phase. One aims at recalcification.

Diet· high calcium, high phosphorus, high protein (meats, fish, skim milk, eggs, green vegetables, fruit juices)

Vitamin D: Drisdol (Winthrop-Stearn) 30 drops daily in milk

Sex hormones: Conjugated estrogen 1.25 mg. (Premarin [Ayerst] oral) and testosterone 10 mg., sublingual, daily. The estrogen is given for 3-week periods with intervals of 1 week.

Osteoblastic Phase. One aims at decalcification.

Diet: A low mineral diet consisting of 0.3 Gm. calcium, 0.5 Gm. phosphorus, and 0.15 Gm. magnesium

Vitamin D: Withheld

Sex hormones: Withheld

Normally on this diet, urinary excretion is 100 to 150 mg. of calcium, 175 to 225 mg. of phosphorus, and 50 to 70 mg. of magnesium. Retention of these elements occurs during the osteoblastic phase.

Magnesium Carbonate: About 4 to 6 Gm. is administered daily, and a qualitative estimation of calcium excretion by the Sulkowitch is sufficient to determine an adequate response.

During the preoperative period in handling fractures, when enforced bed rest imposes the dangers of hypercalcemia, the decalcification regimen is adhered to strictly, the diet consisting of carbohydrates and fats. Intramedullary fixation of the fracture permits immediate ambulation postoperatively, and the calcifying regimen is substituted at this time.

INFANTILE CORTICAL HYPEROSTOSIS (Caffey's Disease[6, 7])

Caffey's disease is a not uncommon idiopathic condition affecting infants less than 6 months of age and characterized clinically by swellings of subperiosteal ossification and constitutional signs of fever, leukocytosis, and an increased sedimentation rate.

Etiology

The cause is unknown. The condition is often confused with hypervitaminosis A. The constitutional signs suggest an infectious origin.

Clinical Picture

The onset is acute. The infant is irritable, fretful and crying. A variable degree of fever is present. Soft tissue swellings appear suddenly over the diaphyses of long bones, par-

[5] Stein, I, Stein, R O, and Beller, M L: Living Bone in Health and Disease, Philadelphia, Lippincott, 1955.

[6] Roske, G: Eine eigenartige Knochenerkrangung im Sanglingsalter, Monatsschr. f. Kinderh. 47:387, 1930

[7] Caffey, J., and Silverman, W. A.: Infantile cortical hyperostosis, Am. J. Roentgenol. 54·1, 1945.



FIG 240. Infantile cortical hyperostosis. The characteristic laminations of subperiosteal new bone are observed about the mandible and the forearm bones.



ticularly the clavicle and the ulna, and over the scapula. Bilateral facial swelling due to involvement of the mandible is common and characteristic. The swelling, although obviously a soft tissue mass, has a ligneous feel, is situated deeply where it apparently is fixed to the underlying bone, is tender but not warm. These swellings precede x-ray changes. When roentgenographic evidence appears, tenderness and fever subside, and swellings are already regressing. The course is protracted with remissions and exacerbations.

Roentgenologic Findings

The diaphyses of long bones, the ribs, the mandible and the scapula are predisposed. The scapular lesion is often unilateral. The typical lesion is the development of a periosteal elevation by an opacity, often laminated (onion-peel) in appearance, representing periosteal new bone formation. The new ossific mass varies in extent, but when it is abundant and extends along the entire shaft it is limited by the attachments of the periosteum to the epiphyseal plate. Periosteal new bone at first exhibits a vague opacity which gradually increases in density and then blends with and thickens the cortex. Later, remodeling restores bone structure and size to normal.

Laboratory Findings

These consist of an increased sedimentation rate, leukocytosis, increased alkaline phosphatase, and anemia which occasionally may require transfusion.

Pathology

Biopsy specimens exhibit periosteal thickening, edema and subperiosteal new bone formation. No evidence of hemorrhage or inflammation is observed.

Differential Diagnosis

The importance of infantile cortical hyperostosis lies in differentiating it from the following conditions:

Hypervitaminosis A. This condition occurs in infants above 12 months of age. The mandible is never involved, fever is absent, a high vitamin A blood level is usual, and discontinuing the vitamin effects a cure in 1 week.

Scurvy. The appearance of subperiosteal ossification is similar. A ground-glass osteo-

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OPOIETIC TISSUES

ertain tumors resemble
in the bone marrow or
The supposition is that
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'presents incomplete dif-

General Characteristics of Hemopoietic Tumors

These neoplasms all have peculiarities common to all:

1. *They involve cancellous bone.* Early, roentgenograms are negative, and symptoms consist of vague "rheumatoid" discomfort, with remissions and exacerbations.

2. *Pain progressively worse and persistent.* Accentuated by a sudden movement or strain. Involvement of vertebrae causes girdle pain (with or without herpes zoster), lower extremity referred pain and abdominal pain

3. *Osteolytic* in hemopoietic tissues especially of vertebrae (results in compression or collapse of bodies), sternum, ribs, pelvis, skull and long bones. Pathologic fractures.

4. *Leukocytosis and secondary anemia.* Involvement of marrow results in premature extrusion of immature cells into the blood stream. With extensive involvement of the blood-forming tissues, leukopenia and severe secondary anemia are typical of the late stage of disease.

5. *Sternal marrow frequently infiltrated.* Sternal puncture is diagnostic.

6. *Sensitive to irradiation.* Relief of pain immediate and dramatic. Ultimate resistance of tumor to irradiation

7. *Enlargement of spleen and lymph nodes*

Malignant Lymphomas

Under this term are included, in descending order of frequency, Hodgkin's disease, lymphosarcoma, reticulum cell sarcoma, and giant follicle lymphoma. The skeleton is involved by direct extension from neighboring lymph nodes, by metastatic spread, or independently within the bone. Common characteristics make it advisable to consider this group with the leukemias.

Hodgkin's Disease

This is a painless progressive involvement of lymph nodes, spleen, liver and skeletal marrow by tissue which is granulomatous in appearance and acts neoplastic.

Clinical Picture

Age. Young adults, usually males

Course. Early swelling of lymph nodes, particularly in cervical area. Nodes at first discrete and painless, later become greatly enlarged and matted together. Severe pruritus,

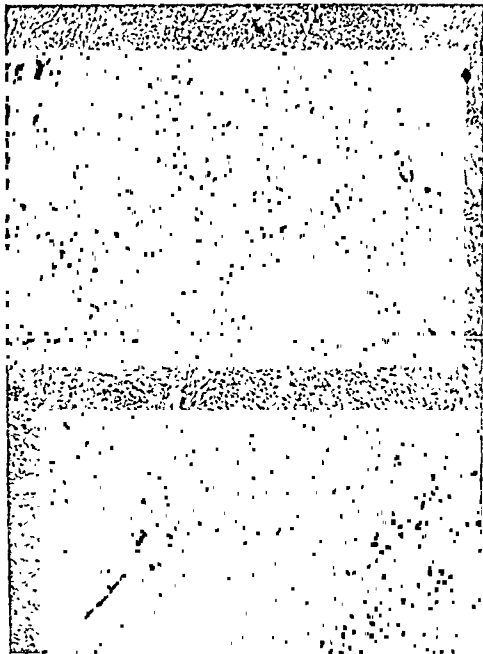


FIG. 241. Sarcoid of bone. Demonstrating rare giant cells, containing typical densely staining material especially in the one at the top of the upper section. Peculiar small accumulations of colloid material. Lymphocytes are sparse and diffusely distributed. Epithelioid accumulations and capillary loops of granulation tissue are seen in the lower photomicrograph. (Turek, S. L.: Sarcoid disease of bone at the ankle, J. Bone & Joint Surg. 35A: 465)

Clinical Picture

Severe involvement throughout the body often is asymptomatic, and the diagnosis is made accidentally. Symptoms, when they do occur, are generally mild and consist of anorexia, malaise and slight elevation of temperature. Lungs and lymph nodes frequently contain extensive infiltrations without symptoms. Skin lesions are common, are usually associated with bone sarcoidosis and present a characteristic appearance They are dusky red, slightly elevated, nonulcerating, nonpruritic nodules, and often are situated over the nose, the cheeks and the ears (lupus pernio).

Bone sarcoid occurs in a high percentage of cases but is asymptomatic except for secondary effects. The phalanges of the hands may contain large sarcoid accumulations which may enlarge and deform the bone but do not invade the soft tissue. When sarcoid granulation tissue causes absorption of subchondral bone, the overlying articular cartilage ineffectually resists joint pressures, and symptoms of degenerative arthritis ensue.

Although the course is prolonged, even for years, spontaneous healing eventually takes place.

Roentgenologic Findings

Irregular, nodular, symmetric densities are

characteristic of pulmonary dissemination. Widening of the mediastinal shadow results from extensive involvement of the hilar lymph nodes.

The picture of osseous lesions depends upon the degree of involvement and concentrations of tubercles in localized areas. The entire medulla may be studded with miliary tubercles without apparent change in bony architecture. On the other hand, multiple, small, circular or oval, punched-out osteolytic areas represent concentrations of granulomatous tissue which replace the bone. No reactive bone formation is observed. The spongiosa is predisposed, but cystlike areas often erode and thin the cortex. The roentgenologic appearance is remarkably similar to gout, but, contrary to the latter, the overlying soft tissues are never swollen.

Laboratory Findings

Variable findings have been reported by different observers. The sedimentation rate is frequently elevated. Hypercalcemia has occurred in cases with extensive bone involvement. Hyperglobulinemia is common. Biopsy of an affected bone or lymph node is diagnostic. In the majority of cases, a negative Mantoux test is significant.

Treatment

No definitive treatment is known. The condition is chronic but benign. In some instances calciferol and dihydrotachysterol have effected rapid resolution of the lesions. In the course of treatment, blood calcium levels are frequently done to avoid the dangers of hyercalcemia.

A painful degenerative arthritis may supervene in a weight-bearing joint adjacent to sarcoid osseous lesions. Arthrodesis may prove to be the most rapid means of rehabilitation. An intramedullary graft may be inserted without fear of destruction by the sarcoid process.

TUMORS OF HEMOPOIETIC TISSUES

The basic cells of certain tumors resemble those normally found in the bone marrow or in lymphoid tissues The supposition is that they arise from a common stem cell, and the ultimate tumor cell represents incomplete differentiation.

General Characteristics of Hemopoietic Tumors

These neoplasms all have peculiarities common to all:

1. *They involve cancellous bone.* Early, roentgenograms are negative, and symptoms consist of vague "rheumatoid" discomfort, with remissions and exacerbations.
2. *Pain progressively worse and persistent.* Accentuated by a sudden movement or strain. Involvement of vertebrae causes girdle pain (with or without herpes zoster), lower extremity referred pain and abdominal pain
3. *Osteolytic* in hemopoietic tissues especially of vertebrae (results in compression or collapse of bodies), sternum, ribs, pelvis, skull and long bones. Pathologic fractures.
4. *Leukocytosis and secondary anemia.* Involvement of marrow results in premature extrusion of immature cells into the blood stream. With extensive involvement of the blood-forming tissues, leukopenia and severe secondary anemia are typical of the late stage of disease.
5. *Sternal marrow frequently infiltrated.* Sternal puncture is diagnostic.
6. *Sensitive to irradiation.* Relief of pain immediate and dramatic. Ultimate resistance of tumor to irradiation
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Malignant Lymphomas

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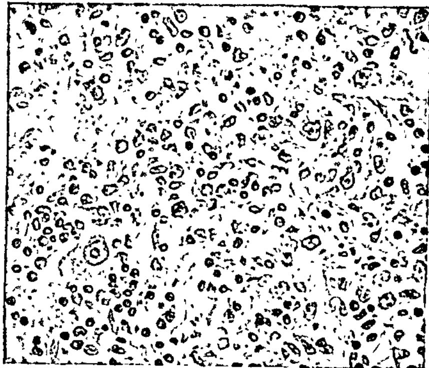


PLATE 40. Hodgkin's disease (× 415). Typical Reed-Sternberg cells are shown.

weakness and weight loss. Liver and spleen enlarged and palpable. Initial bony involvement of the marrow is painless. Slowly, bone pains, at first rheumatic in type and intermittent, become more severe and persistent. Tender swellings appear over the bones, particularly over the ribs and the sternum. Vertebrae frequently involved with a high incidence of compression fractures. Compression of intraspinal structures is responsible for referred chest and lower extremity pain. Eventually, compression and direct invasion of vital structures lead to a fatal termination.

BONE INVOLVEMENT. Wherever red bone marrow exists, particularly vertebrae, ribs, sternum, pelvis, skull, scapula, and the metaphyseal area of long bones.

PATHOLOGIC FRACTURES are rare except for multiple vertebral compression

FINDINGS The superficial lymph nodes are enlarged and discrete early, very large, tender, and matted together late. Late in the course of the disease, the bones are tender and superficial swellings may be palpable. Variable fever.

LABORATORY. High leukocytosis and secondary anemia. Eosinophilia. Lymph node biopsy diagnostic. Alkaline phosphatase elevated with bone involvement.

Roentgenologic Findings. Early negative. Later osteolytic and diffuse Occasionally, some medullary reactive bone density about the lesions may be seen if the growth is slow. Superficial cortex indentation represents pressure erosion.

Pathology. GROSS. The disease frequently starts in the cervical lymph nodes and spreads to axillary, mediastinal, retroperitoneal, mesenteric and inguinal lymph node groups. The nodes remain discrete at first, even though enlarged. On cut section they are pink, homogeneous, and elastic or firm. Later, the glands are further enlarged, grayish white, firm and matted together in a mass of fibrous tissue. Where a mass of glands overlies a bone, the latter undergoes localized erosion. Within the bone marrow, the granulomatous tissue is seen in discrete or confluent masses eroding the cortex from within. The tissue may extend to the subperiosteal area elevating the periosteum with minimal reactive bone. The granulomatous tissue in vertebrae may extensively replace the marrow with consequent collapse of the body and extend to the peridural space with compression of nerve structures. The disks are never involved.

MICROSCOPIC. In a reticular stromal network is seen a variety of cells including endothelial cells, plasma cells, lymphocytes and eosinophils. The characteristic cell is the Reed-Sternberg cell, a large endothelial cell with multilobed nuclei. Where the tumor tissue encroaches upon the bone trabeculae, the latter is eroded or absorbed without the benefit of

osteoclasts. Early, the tissue is granulomatous. Later, a highly cellular tissue in which the cells are more uniform and round becomes pervaded with fibrous elements.[9, 10]

Treatment. Palliative treatment consists of irradiation to the bones and the lymph nodes. From 600-1500 r will relieve pain at a clinically manifest bone lesion and may relieve neurologic complaints due to central nervous system pressure. Radioactive calcium and intravenous nitrogen mustard are now being used with questionable results.

LYMPHOSARCOMA

Lymphosarcoma of bone is very rare. When it occurs, multiple bone involvement is the rule The tumor extends diffusely throughout the bone and is centrally destructive without formation of reactive bone. Clinically, pain occurs early and, like other myelomatous lesions, the vertebrae are commonly involved, producing referred girdle chest discomfort and lower extremity pain Laboratory examinations reveal leukocytosis and relative lymphocytosis, eosinophilia and secondary anemia. As the marrow is replaced more extensively, severe anemia and leukopenia are late manifestations. Pathologic fractures occur. Eventually, the lymph nodes are involved. The roentgenogram may show a long bone diffusely osteolytic and *not surrounded by reactive bone.* Vertebrae are partially collapsed. Rarely, the lesion may excite osteogenesis. The tumor is seen to extend diffusely throughout the medulla, eroding the cortex and extending through the haversian canals to the subperiosteal space. The tumor is rubbery and opaque. Microscopically, a diffuse growth of lymphocytes in a reticular tissue stroma is seen. Multinucleated cells are not observed Treatment consists of irradiation, which promptly relieves pain The outlook is hopeless, exodus occurring within 3 years [10-12]

[9] Geschickter, C. F, and Copeland, M. M : Tumors of Bone, Philadelphia, Lippincott, 1949

[10] Jaffe, H L Skeletal manifestations of leukemia and malignant lymphomas, Bull Hosp. Joint Dis 13 217, 1952.

[11] Baldridge, C. W, and Awe, C D.: Lymphosarcoma—a study of 150 cases, Arch Int. Med 45·161, 1930.

[12] Craver, L. F, and Copeland, M M : Lymphosarcoma in bone, Arch Surg 28 809, 1934.

THE LEUKEMIAS

Leukemia is a condition in which an overproduction of immature forms of lymphocytes (lymphocytic leukemia) or polymorphs (myelocytic leukemia) results in extensive replacement of other elements of the hemopoietic and lymphoid tissues. It occurs as an acute or chronic form, the former being most common and predominantly in infancy and childhood. The clinical picture includes fever, anemia, enlarged lymph nodes, splenomegaly, hepatomegaly, tender bones and swollen painful joints, and progressive weight loss. Laboratory examination reveals immature white blood cells in the blood stream regardless of whether a leukocytosis or a leukopenia (the aleukemic form) exists. Thrombocytopenia leads to increased bleeding time and purpuric tendencies. Symptoms include asthenia and bone and joint pains, thereby simulating acute rheumatic fever. A slightly tender swelling may be palpable over the superficially accessible bones as the ulna. Estimates of bone involvement run as high as 50 per cent and usually is a late manifestation.[10, 13-15]

The leukemic tissue infiltrates and replaces the bone marrow diffusely throughout the bone, resorbing the cancellous and later the cortical bone. It reaches and elevates the periosteum, which reacts to form thin laminations of reactive bone parallel with the shaft. Pathologic fracture occurs chiefly in the ribs. Microscopically, the cells of lymphatic leukemia produce a picture similar to lymphosarcoma. The cells of myeloid leukemia are immature myeloblasts and myelocytes.

Roentgenologic findings consist of *diffuse osteoporosis, punctate rarefied areas* throughout the cortex producing a ragged appearance, a transverse area of increased *radiolucency on the metaphyseal side of the epiphyseal line,* and thin *laminations of periosteal reactive bone.* The picture is mainly one of osteolysis, but in chronic leukemia in the adult, osteosclerosis throughout the skeleton is common.

Treatment. Irradiation of the bones is effec-

[13] Hoxie, T B : Bone and joint pain in leukemia, New England J. Med 238.733, 1948.

[14] Dresner, D : Bone and joint lesions in acute leukemia and their response to folic acid antagonists, Quart. J. Med. 19.339, 1950

[15] Dale, J. H, Jr.. Leukemia in childhood, J. Pediat. 34.421, 1949.

tive in relieving bone pain early in the course of the disease. Later, its effectiveness is lost. The use of antifolic acid substances, radioisotopes, ACTH and cortisone may be tried. The condition is usually fatal with the advent of a severe hemorrhagic complication.

It is important to recognize the fact that leukemic infiltrates in the synovium of joints are a common manifestation of the disease, and differentiation by biopsy is necessary.

Chloroma

This is a form of lymphatic or myeloid leukemia in which bone manifestations form a prominent feature. Characteristically, the tissue contains a greenish pigment so that the involved tissue, whether bone, lymph gland, or spleen, appears green on cut section. Children near puberty are affected, and the condition is rapidly fatal within 5 months. The *greenish tumors* most commonly invade the skull and produce symptoms referable to the structures about the head. Typically, the growth extends within the orbital cavities and causes an *exophthalmic protrusion* and edema of the eyelids. Other bones with much red marrow are also involved, and the tissue may even extend beyond the bone and infiltrate surrounding muscles and tendons. Clinical, x-ray, and microscopic pictures are similar to other forms of leukemia. However, the extremely malignant character of the tissue is reflected in the large atypical monocytes, the premature myeloblasts, with numerous mitoses and hyperchromatic nuclei. Treatment is confined to small doses of x-rays, but the outlook is hopeless.

Reticuloendotheliosis

All lymphoid and myeloid tissue contains a mesh of reticular tissue in which are the *primitive reticular cells.* These primitive reticular cells contain pale oval nuclei and protoplasmic prolongations which join those of neighboring cells The reticular fibers are strewn throughout the protoplasmic network. The primitive reticular cells transform into all types of blood and connective tissue cells. They frequently change into similar appearing but larger cells with abundant cytoplasm, large pale nucleus, and spindle shape.

These are the *fixed macrophages* which engulf dead cells and debris and are capable of ingesting certain dyes, such as lithium carmine. The inclusions of the macrophages stain deeply with neutral red. These macrophages are flattened and help to form the walls of sinuses or channels through which lymph flows and therefore are thought of as endothelium. However, endothelial cells are incapable of phagocytosis and cannot be stained with lithium carmine. Under certain conditions, as when foreign matter is present, the fixed macrophages *transform into free macrophages and* float free in the lymph.

The primitive reticular cells also develop into *lymphocytes* and *myelocytes.* The lymphocytes vary in size and contain a large darkly staining nucleus and a scanty basophilic cytoplasm which contains no inclusions.

Eosinophile cells are normally found in the connective tissue in certain situations as the mammary gland. Under pathologic conditions, they may migrate from the blood stream and settle in large numbers in the connective tissue.

Plasma cells are transformed from lymphocytes and apparently constitute the ultimate *in differentiation as they are unable to transform* into any other cell.

Fat cells are found in variable number in loose connective tissue, particularly about vessels at the site of the primitive reticular cells In development of the fat cell, multiple small droplets accumulate before fusing into one large drop. The original primitive cell loses its elongated processes and becomes more polyhedral before rounding up.

In formation of *foam cells* hereafter described it is tempting to believe that these are transitional fat cells containing certain lipoid substances other than neutral fat.

This foregoing description of reticuloendothelial histogenesis suggests a common denominator for the reticuloendothelioses which include· (1) Hand-Schuller-Christian disease; (2) eosinophilic granuloma; (3) Letterer-Siwe's disease, (4) Gaucher's disease; and (5) Niemann-Pick's disease. Many observers refer to them as variants of the same basic disorder.[16, 17]

[16] Hodgson, J R, Kennedy, R J. L, and Camp, J D Reticuloendotheliosis, Radiology 57:642, 1951

[17] Ponseti, I Bone lesions in eosinophilic granuloma, Hand-Schuller-Christian disease, and Letterer-Siwe disease, J Bone & Joint Surg 30A·811, 1948.



FIG. 242. Eosinophilic granuloma of body of T5 vertebra. Marked deossification and beginning collapse. Preservation of disk spaces and adjacent vertebrae distinguishes this from tuberculosis.



FIG. 243. Eosinophilic granuloma, involving a vertebral body. The body of the 5th thoracic vertebra is flattened. Pathology proved by biopsy. After healing of the lesion, growth of the body is resumed; if a sufficient growth period remains, the body may regain much of its original size. This may explain what has been described as Calvé's disease of the spine.

Histologically, the pathology consists of formation of tissue composed of variable amounts of proliferated reticuloendothelial cells, macrophages containing debris and fat, foam cells, eosinophils, lymphocytes, plasma cells and, about areas of necrosis, multinucleated giant cells. Involvement of the skeleton by reticuloendothelial granuloma causes destruction, in single or multiple localized areas, without reactive new bone formation. The tissue is usually soft and friable and varies from reddish (vascular) and yellowish (lipoid) to grayish (fibrous) and firm. Roentgen findings consist of areas of rarefaction, multiple or solitary, involving one or more bones. Skull involvement is frequent. Lesions are seen in other flat bones (pelvis, scapula), and vertebral lesions are rare. The destruction may be large and display sharply scalloped edges. Clinically, lymphadenopathy, splenomegaly, hepatomegaly and bone lesions are common denominators The diagnosis is established by biopsy.

The macrophages of certain reticuloendothelioses contain specific lipoid substances. These so-called *lipoid histiocytoses* include Hand-Schüller-Christian disease, Gaucher's splenomegaly and Niemann-Pick disease. When lipoid inclusions are not a prominent feature, these *nonlipoid histiocytoses* are eosinophilic granuloma and Letterer-Siwe disease.

EOSINOPHILIC GRANULOMA

This is a solitary benign bone-destructive

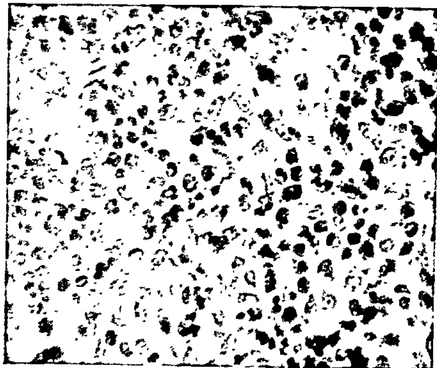


PLATE 41. Eosinophilic granuloma. Note, in addition to the eosinophils, macrophages which are large, pale, rounded bodies with a vesicular nucleus and abundant cytoplasm with indistinct margins. (× 570)

lesion characterized by the presence of eosinophils.[18, 19]

Clinical Picture

AGE. Children or young adults

AREAS OF PREDILECTION. Ribs, flat bones. vertebrae

SYMPTOMS. Relatively acute onset, pain

FINDINGS. Swelling over bone, tenderness, low grade fever, leukocytosis. *Solitary lesion.* Rarely multiple. Eosinophilia

COURSE. Acute, over a few weeks, tendency to spontaneous healing

PATHOLOGIC FRACTURES. Common. Vertebral collapse.

Roentgenologic Findings. Well-localized radiotranslucent area without reactive bone formation. Cortex destroyed and expanded as a shell, perhaps with slight periosteal new bone

Pathology. GROSS. Soft friable reddish-gray or reddish-yellow tissue within a bone defect. Healing lesion exhibits the firm gray fibrous tissue with bone formation.

MICROSCOPIC. The tissue is a very cellular granulomatous one in which eosinophils and macrophages predominate, the stroma being reticular. The macrophages are large, mononuclear, and contain blood pigment. The stromal cytoplasm is foamy, but foam cells, lymphocytes, and plasma cells are in the minority. As the lesion heals, the eosinophils disappear, the foam cells become more numerous, and finally the fibrous proliferation supervenes. Ossification completes the process. Multinucleated giant cells appear only about areas of hemorrhage and necrosis and are not a diagnostic feature.

Diagnosis. Biopsy is necessary. The roentgenographic appearance resembles metastatic tumors, multiple myeloma and other lesions.

Treatment. The lesion is radiosensitive. Curettage and filling the defect with bone chips obtains a rapid cure. It should be supplemented with x-ray therapy. Inaccessible lesions are treated with irradiation. The prognosis is good.

Vertebra Plana Due to Eosinophilic Granuloma. Calvé in 1924 described what he called osteochondritis of the vertebral body. This consisted of the rapid development, usually in a child, of an osteolytic process in a single vertebral body resulting in marked flattening and increased density, the contour appearing like the edge of a coin or a disk, and not wedge-shaped. Clinically, the onset is gradual or rapid, then rapid progression. Back pain, muscle spasm, limited back motion, night cries and a prominent spinous process or gibbus

[18] Hatcher, C. H.: Eosinophilic granuloma of bone, Arch. Path. 30:828, 1940.

[19] Jaffe, H. L., and Lichtenstein, L.: Eosinophilic granuloma of bone, Arch. Path. 37:99, 1944.

form the characteristic picture. Spontaneous reossification with variable loss in height of the body occurs over several years. Recently, several identical cases have been described in which eosinophilic granuloma proved to be the causative condition.[20] Treatment consisted of bed rest, splinting and irradiation.

Letterer-Siwe Disease

This is an acute generalized reticuloendothelial disease occurring in infants and invariably fatal.

Clinical Picture

Age. Below 3

Areas of Predilection. Sites of red marrow, i.e., flat bones, vertebrae, skull, etc.

Symptoms and Course. Usually ushered in with fever and infection of respiratory, gastrointestinal, or genito-urinary systems. Generalized lymph node swellings at first followed by splenomegaly and hepatomegaly. Skin lesions, purpura, gingival inflammation and ulceration, hemorrhages, progressive anemia, thrombocytopenia, aplastic anemia and death. Destructive lesions in bones. Fatal in a few years

Pathology. Grossly, grayish-yellow nodules replace cancellous and cortical bone.

Microscopically, marked proliferation of the reticulum cells and predominance of large mononuclear cells with vacuolated cytoplasm are seen. The latter are histiocytes, or macrophages, which display large dense malignant-appearing nuclei. Foam cells are rare or nonexistent. This lack of lipoid classifies this as a nonlipoid histiocytosis.

Treatment. There is no known beneficial treatment.

The following conditions are known as the lipoid histiocytoses or the xanthomatoses. They are characterized by the deposit of specific lipoid substances within so-called *foam cells*

Hand-Schüller-Christian Disease

This is a congenital reticuloendothelial tissue disease which characteristically produces large destructive lesions of bone, particularly in the skull.

[20] Compere, E L, Johnson, W. E, and Coventry, M. B Vertebra plana (Calvé's disease) due to eosinophilic granuloma, J Bone & Joint Surg. 36A·969, 1954



Fig. 244. Eosinophilic granuloma, involving the shaft of the radius. The lesion is osteolytic without reactive bone formation. In this case the growth has been resected, and a cast applied. A sheath of periosteal new bone is beginning to form. (Dr. Clinton Compere's case)

Clinical Picture

Age. Begins in childhood and progresses into adulthood.

Areas of Predisposition. Membranous bones. particularly the skull. Less commonly the long bones

Symptoms. Characteristic triad: (1) defects in skull, (2) diabetes insipidus and (3) exophthalmos. Splenomegaly, skin pigmentation, endocrine disturbances and occasional elevation of the blood cholesterol

Course. Chronic. May last many years

Roentgenologic Findings. Very large well-demarcated defects in the skull, other flat bones and long bones. Cortex is thinned. No periosteal reaction. Multiple defects

Pathology. The neoplastic tissue destroys and replaces bone and extends out into the soft tissue. Within the orbital cavity it pushes the eyeball forward to cause exophthalmos. In the pituitary fossa, it destroys the pituitary gland, causing sexual and growth disturbances (anterior pituitary), then extends to the posterior pituitary and the hypothalamus to produce diabetes insipidus.

Microscopically, the tissue consists of a very cellular reticuloendothelial tissue enmeshing great numbers of macrophages, eosinophils,

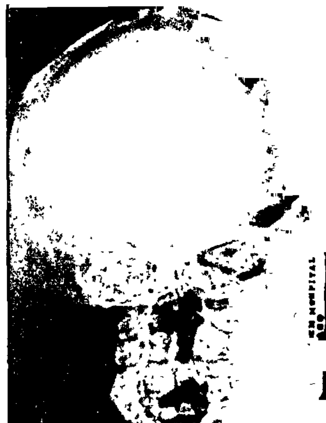


Fig. 245. Hand-Schüller-Christian disease, showing sharply demarcated osteolytic skull lesions.



plasma cells and lymphocytes. The macrophages are the characteristic xanthoma foreign body giant cells These are large cells, often binucleated, laden with cholesterol lipoid and blood pigment and resembling Reed-Sternberg cells. They are more common in the lymph nodes and the spleen. Foam cells may or may not be present

Treatment. The lesions are radiosensitive The bones may completely reossify following such treatment. Diabetes insipidus is controlled by posterior pituitary extract. Growth and sexual disturbance may respond to anterior pituitary extract.

"Histiocytosis X"

Eosinophilic granuloma, Letterer-Siwe disease and the Hand-Schüller-Christian syndrome are regarded by many observers as manifestations of the same pathologic entity.[21] The common denominator is an inflammatory histiocytosis.

The histiocyte or free macrophage is a large cell with outstretched ragged outlines containing a large oval or kidney-shaped nucleus containing coarse chromatin granules and possessing cytoplasm which is often vacuolated. Characteristically, it will take up acid dyes such as trypan blue and lithium carmine. Under conditions of inflammation, histiocytes become abundant and round.

Eosinophilic granuloma, Letterer-Siwe disease and Hand-Schüller-Christian syndrome, hereinafter described as "E-G," "L-S" and "S-C," are characterized by an intense accumulation of histiocytes. L-S represents the acute (or subacute), and S-C the chronic, disseminated forms of the same malady. The accumulation of eosinophils represents a rapidly developing defense to the same etiologic agent, whatever it may be.

The terms "reticuloendotheliosis" or "reticuloendothelial hyperplasia" are regarded as improper designations, because hyperplasia of reticuloendothelial cells may occur in neoplasms, response to abnormal lipid storage and inflammatory conditions Further, the typical cells in question are histiocytes displaying phagocytosis.

The localized form of the disease is expressed as an eosinophilic granuloma. One or several, occasionally many, lesions may be observed, no constitutional illness occurs, and cutaneous, pulmonary or other extraskeletal involvement is absent. The outcome is favorable. The lesion is destructive, develops rapidly, breaks through the cortex and heals rapidly after curettement or irradiation. It

[21] Lichtenstein, L.: Histiocytosis X, Arch Path 56 84, 1953

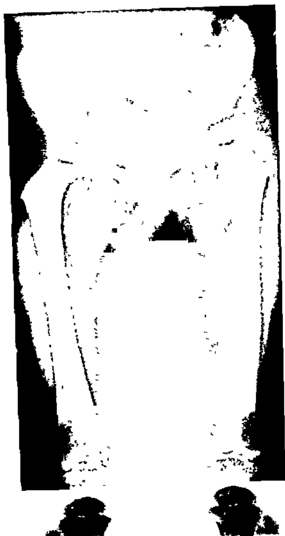


Fig. 246. "Histiocytosis X" in a 2-year-old female in femur and ischium. Central destruction also involving cortex and little reactive bone formation, except some periosteal ossification. Punched-out lesions also in skull and rib. Biopsy confirmed diagnosis. Rapid reossification occurred under the influence of x-ray therapy.

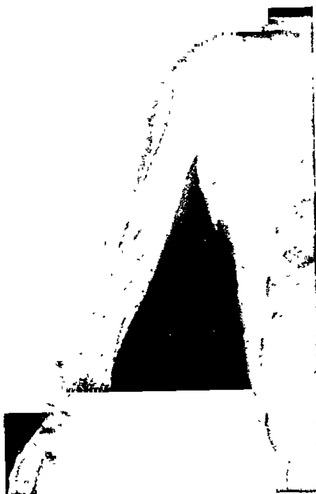


Fig. 247. Osseous lesion of histocytosis, healing of pathologic fracture.

may heal spontaneously. It represents the most successful defense against the disease.

On the other hand, eosinophilic granuloma may represent a phase of the disease. Dissemination may already have occurred or may become manifest later in other parts of the skeleton or the viscera. Therefore, when an isolated lesion of eosinophilic granuloma is observed, one must reserve judgment for months or years. Eventually, evidence of dissemination is revealed by fatigability, chronic malnutrition, weight loss, slight fever, predisposition to infection, and other skeletal defects. Then the condition may be classified as S-C or L-S.

There is great variation in clinical severity. Even subacute disseminated histiocytosis may slow its course, become chronic and undergo clinical remission. The subacute case with a fatal termination is not necessarily restricted to infants and young children. The S-C type, although protracted over many years, may prove to be fatal from extensive pituitary damage or serious pulmonary infiltration and fibrosis.

Pathologic Similarities. An intense inflammatory histiocytosis develops at first, with or without eosinophilic reaction. A profusion of eosinophils, although it frequently occurs as an isolated lesion, may develop at any time in the course of an acute (L-S) or chronic (S-C) disseminated histiocytosis. It is revealed roentgenologically as a rapidly destructive lesion and may persist for a year or more. The eosinophils disappear, and histiocytes ac-



FIG. 248. Gaucher's disease. The cortex of the shaft is thinned, and the medullary cavity is widened because of replacement of cancellous bone by pathologic tissue.



FIG. 249. Gaucher's disease, showing the characteristic Erlenmeyer flask appearance (Fischer sign) at the lower end of the femur. Diagnostic findings in roentgenograms include a generalized mottled appearance, increased breadth of the marrow cavity, thinning of cortices, and flaring of distal part of the femur. (Kroboth, F. J., Jr., and Johnson, E. W., Jr.: Osseous Gaucher's disease, S. Clin. North America 32: 1141)

cumulate and are distended by lipoid deposits. These lipophages have a foamy appearance and give a distinctly yellow color to the tissue. The lesion then has a tendency to undergo fibrosis

In both subacute and chronic forms of the disease, skeletal involvement is diffuse, and extraskeletal sites are numerous. The latter are most often recognized as papular eruptions and ulcerations of superficial mucocutaneous surfaces, lymph node enlargements and pulmonary infiltrations. A pathognomonic finding in chronic disseminated histiocytosis because of extensive pulmonary infiltrations and fibrosis is the "honeycombing" seen on the roentgenogram. Episodes of spontaneous pneumothorax are common.

GAUCHER'S DISEASE
(Gaucher's Splenomegaly)

Gaucher's disease is a congenital reticuloendothelial disease characterized by enlargement of the spleen to enormous size and deposit of the specific cerebroside, kerasin, in all lymphoid tissue.[22-25]

Clinical Picture

AGE. Infantile form starts before 6 months and becomes fatal before 2 years. Adult form starts in late childhood or adolescence, relatively benign, lasts indefinitely.

SYMPTOMS AND FINDINGS. Infantile form has acute onset, adult more common form has insidious onset. Involvement and enlargement of all lymphoid tissues as lymph glands, liver, thymus, tonsils, and especially the spleen. Symptoms of rheumatic pains in the long bones, and findings of low-grade fever, yellowish-brown skin pigmentation, splenomegaly, anemia and leukopenia. Thrombocytopenia and hemorrhagic diathesis are common.

PATHOLOGIC FRACTURE is common.

STERNAL PUNCTURE and chemical identification of the lipoid establish the diagnosis

[22] Aballi, A. J, and Kato, K.: Gaucher's disease in early infancy, J. Pediat 13:364, 1938

[23] Kato, K: Changes of bone in Gaucher's disease, Tr. Am. Pediatric Soc. 43:43, 1931.

[24] Gaucher, P. C E: De l'Epithelioma Primitif de la Rate, These de Paris, 1882.

[25] Gordon, G. L: Osseous Gaucher's disease, Am J. Med 8.332, 1950

Roentgenologic Findings. Multiple areas of *rarefaction* are strewn throughout the bones, giving them a moth-eaten or ragged appearance. Characteristically, the process grows extensively at the metaphyses of long bones, particularly the femur, where the cortices are thinned and wavy, and distended to form a *bulbous or flasklike contour of the bone.* Extreme destruction of the bone and *pathologic fracture* are not unusual. Femurs, vertebrae, and sternum are most commonly involved.

Pathology. GROSSLY, one finds enlargement of lymph nodes, spleen and liver. Cut section of these tissues shows multiple areas of white and pale-yellow nodules. The skeleton is extensively destroyed by the tissue, especially the vertebral bodies, which may become osteoporotic and compressed. The bone is resorbed and replaced by numerous white or pale-yellow nodules. The end of a long bone may be thoroughly destroyed, and the neoplastic tissue may invade the joint structures. The skull is seldom involved.

MICROSCOPICALLY, extensive sheets of *characteristic large pale phagocytes* are seen. These possess a large vesicular nucleus and a finely granular or refractile *cytoplasm containing the lipoid, kerasin.* The cells are closely packed, but occasional areas of lymphocytes and the reticular stroma are visible The histology resembles that of Niemann-Pick disease, but the latter's characteristic cell contains vacuoles.

Treatment. Irradiation to the affected bones and viscera is of questionable benefit. Splenectomy may effect symptomatic improvement but no cure. If x-ray treatment is undertaken, caution is imperative because of the leukopenia and the thrombocytopenia already present.

NIEMANN-PICK DISEASE

This is a rare form of lipoid disease in the infant. Clinically and pathologically, it closely resembles the acute form of Gaucher's disease in infants. However, it is distinguished by storage of the specific lipoid phospholipid which is also called sphingomyelin. A strongly yellow color is imparted to the enlarged liver, the spleen, the lymph nodes and bone marrow. The course is so rapidly fatal that destructive lesions of bone are a rarity. The disease is fatal by lung and central nervous system involvement in the second year.



FIG. 250. Osseous Gaucher's disease. The bony trabeculae are thick and partially necrotic. The marrow spaces are filled with compact collections of Gaucher's cells which are large and polyhedral and contain small nuclei and abundant pale, finely granular cytoplasm. (× 350) (Gordon, G. L.: Am. J. Med. 8:332)

RETICULUM CELL SARCOMA

This is described in the section on Bone Tumors.

TUMORS INVADING BONE FROM OVERLYING STRUCTURES

These include neurogenic sarcoma, myosarcoma and liposarcoma.

NEUROGENIC SARCOMA (Malignant Schwannoma; Malignant Neurinoma)

The tumor originates from the nerve sheath as shown by the microscopic appearance: wavy nuclei, wirelike fibrillae, palisading, myxomatous intercellular substance and tumor giant cells. It resembles fibrosarcoma in the fibrillar structure, and the varying degrees of malignity as seen in the spindle cells. Ewing be-

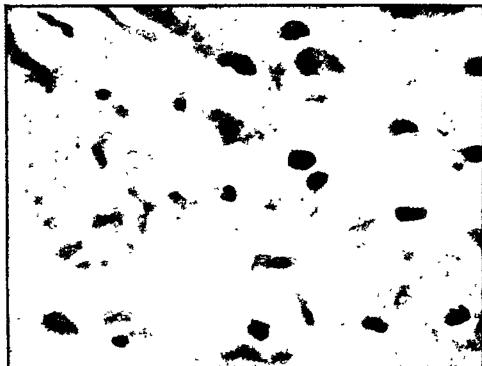


PLATE 42. Rabdomyosarcoma (× 1,200, oil immersion). The typical cells, including the tandem and racquet types, are well displayed. One should seek transverse striations as evidence of origin from muscle.

lieves that the majority of fibrosarcomas of soft-tissue origin are neurogenic in origin. Symptoms of nerve involvement as pain and tingling are common.

Pathology. GROSS. The tumor is a soft, beefy red, semitranslucent fleshy tissue or may be jellylike and translucent. It extends along a nerve trunk as multiple lobulated tumors and infiltrates as a gray translucent material in the soft tissues. It destroys the adjacent cortex, spreads throughout the medullary cavity and may even extend into the joint.

MICROSCOPIC. Tightly packed elongated spindle cells are lined up in parallel rows, resembling a fibrosarcoma. However, these spindle cells are longer, darker and are rippled or waved. Myxomatous areas containing glial-like elements and degenerating cells are frequent. Around the myxomatous areas are seen elongated nuclei in cells of the neurilemmoma type in a palisade arrangement, more commonly seen in tumors less malignant The more malignant tumor displays large pleomorphic nuclei, spindle cells so tightly packed as to crowd out the myxomatous tissue, bizarre tumor giant cells, and relative absence of palisading and whorls.

Prognosis and Treatment. The outlook is very poor. Attempts at local excision are doomed to failure. Even though the tumor microscopically appears benign, radical amputation and deep x-ray therapy are justified.

LIPOSARCOMA

This tumor is very rare. It occurs wherever there is fat; consequently, it may originate not only external to the bone but also within the medullary cavity of a bone. It is very malignant and destructive to bone. Therefore, an intra-osseous tumor rapidly extends into the soft tissues, and an extra-osseous tumor easily invades the bone. Microscopically, the common cells are spindle cells and polyhedral cells with granular cytoplasm containing fat droplets. These pale swollen polyhedral cells resemble those seen in a hypernephroma. Cells resembling fetal fat cells and tumor giant cells may be present. A common extra-osseous location is in the intermuscular tissue about the knee joint. Grossly, the tumor is lobulated and soft and on cut section shows loose fatty and myxomatous areas. It metastasizes quickly to the lungs. In the intra-osseous type, on the other hand, spread is delayed, and metastases occur first to other bones and finally to the lungs. Liposarcoma primary in bone is discussed in the section on Bone Tumors.

RHABDOMYOSARCOMA

This rare tumor is composed of striated muscle, and it is very malignant. Although its usual situation is in the heart, the bladder, the vagina and the cervix, it may occur in voluntary muscle where it secondarily destroys and

invades the bone. Grossly, the tumor may be only a few inches in diameter and irregularly ovoid in shape. Although appearing encapsulated, infiltration beyond the capsule is demonstrable. The tumor is generally firm and elastic and on cut section contains small areas of hemorrhage and necrosis. Microscopically, a wide variety of pleomorphic cells include round, spindle, droplet or "racquet," and tandem cells plus giant cells are seen. The nuclei are large, and the cytoplasm is deeply acidophilic. Many cells contain cross striations characteristic of skeletal muscle. The recommended treatment is wide excision or amputation plus deep x-ray therapy. The outlook is very poor.

18

Peripheral Vascular Disease

The subject of vascular disease overlaps many other fields of medicine and surgery. A study of blood vessels of the extremities, although closely allied to the field of orthopaedic surgery, cannot be considered apart from the rest of the body. The state of the heart and the large vessels in the chest and the abdomen may have a direct relationship to circulatory disease of the extremities. For example, a subacute bacterial endocarditis may be the cause of embolic phenomena; occlusion at the bifurcation of the aorta results in arterial insufficiency in the lower extremities. The orthopaedic surgeon must feel free to consult with the internist and the general surgeon

DIAGNOSIS OF VASCULAR DISEASES OF THE EXTREMITIES[1]

The orthopaedic surgeon must have a rudimentary knowledge of symptoms and objective manifestations caused by various vascular disturbances. By following a definite plan of eliciting the history, examining the affected parts and performing special tests, a comprehensive examination may be completed. The following pertinent points must be investigated.

Pain

This is the commonest symptom. It is either persistent or intermittent.

Persistent pain is due to a number of causes:

1. *Pain of Ulceration and Gangrene and Pretrophic Pain.* Ulceration and gangrene in thromboangiitis obliterans causes extremely severe pain. In arteriosclerosis obliterans such pain is of lesser severity Pain is partially relieved by the dependent position and heat; it is intensified by elevation and cold. The patient characteristically sleeps with the legs hanging over the side of the bed. Similar severe pain, due to ischemia, is known as "pretrophic pain" because it is the forerunner of ulceration and gangrene.

2. *Pain of Sudden Arterial Occlusion.* The onset of excruciating pain may be sudden or gradual, reaching its maximum intensity in several hours. Numbness, coldness and tingling are often associated.

3. *Pain of Ischemic Neuritis.* Pain is severe, diffuse, spasmodic and does not correspond to distribution of peripheral nerves. The character of the discomfort is sharp, shooting; or pulling, tearing, agonizing; or burning, throbbing. During paroxysms, the extremity may become mottled, dark and bluish red owing to excessive vasoconstriction. Between paroxysms, a constant, dull, diffuse, shifting ache is present. Paroxysms occur most often at night and may last for several hours.

4. *Pain of Arteritis, Phlebitis and Lymphangitis.* Acute arteritis causes mild pain, and the artery is tender to pressure. Chronic arteritis is painless Phlebitis causes mild pain and tenderness of the involved vein. Lymphangitis is evidenced by a red line, soreness and tenderness.

Intermittent pain is due to the following:

1. *Temperature Changes.* Exposure to cold when it causes symptoms of Raynaud's disease (numbness and stiffness during stage of pallor and cyanosis, tingling in stage of rubor) seldom causes pain. When Raynaud's phenomenon is associated with pain, a predisposing condition must be suspected, e.g., cervical rib, peripheral neuritis, thromboangiitis obliterans

Pain of erythermalgia is provoked by warmth, typically is a burning distress and affects the ball of the foot, the tips of the toes or corresponding parts of the hand. It is relieved by cold.

[1] Allen, E V, Barker, N W, and Hines, E A, Jr Peripheral Vascular Diseases, Philadelphia, Saunders, 1948.

2. *Exercise.* Intermittent claudication occurs almost exclusively in chronic occlusive arterial disease. It is dependent on deficient circulation in contracting muscles. It is a cramp which occurs with exercise. Rest will relieve the severe pain, but soreness, muscle tenderness and fatigue persist for some time. As occlusion and diminished blood flow is greater in degree, the amount of exercise required to bring about pain is less. The site of claudication indicates the level of occlusion. For example, claudication of the arch of the foot suggests occlusion at or above the ankle. Lessening of intensity and frequency of claudication suggests establishment of collateral circulation. The presence of arterial pulsations in a vessel affected by intermittent claudication suggests a vasospastic tendency, but organic vascular disease eventually becomes apparent. Intermittent claudication has been explained as an abnormal accumulation of a metabolic substance, "factor P," in muscle.

Quantitative Measurement: The patient walks at a rate of 120 steps per minute. The time elapsing between the beginning of the test and the occurrence of pain is known as "claudication time."

3. *Posture.* Chronic venous insufficiency causes an ache or a feeling of heaviness after prolonged standing. It is lessened by walking and relieved by recumbency.

Cramps in the legs occurring while in bed are not a part of organic occlusive arterial disease. The cause is unknown They may arise when stretching or may appear in the course of sleep.

Color Changes

The skin color reflects (1) the amount of blood and (2) the color of blood in the minute vessels of the skin. The more slowly blood flows, the more oxygen it gives up and the more cyanotic the skin becomes. Warmth increases the rate of dissociation of oxygen from the blood; cold has the opposite effect.

When blood flow is rapid, minute vessel tone will be high, and the skin will be warm and pale pink. A warm, deeply colored red skin is due to vasodilatation effected by inflammation, reflexly or by drugs.

The following are typical clinical examples.

Raynaud's Disease. Pallor (cessation of blood flow), cyanosis (stagnation of blood in capillaries), rubor (excessive blood flow). These are produced respectively by arteriolar constriction, capillary dilatation and arteriolar plus capillary dilatation.

Thrombophlebitis. Cyanosis (stasis of blood). Due to obstruction to outflow of blood.

Sudden Arterial Occlusion. Pallor (absence of blood). Due to spasm of main vessel and collaterals.

Erythermalgia. Red warm skin is due to arteriolar and capillary vasodilatation.

Chronic Occlusive Arterial Disease. Pallor on elevation (insufficient blood); the blood pressure cannot overcome circulatory obstruction plus the effect of gravity. Cyanosis on dependency (slowed blood flow); the minute vessels are chronically dilated as a result of ischemia.

Normally, elevation of an extremity above the level of the body will cause a pallor, and the normal color returns within 10 seconds after the part is returned to a dependent position. When arterial circulation is impaired, the pallor is extreme in degree and, if arterial occlusion is irregularly distributed, patchy; on lowering the extremity the color, which is a rubor or cyanotic redness, returns very slowly and often in an irregular or patchy manner.

Sensitivity to exposure to cold, as detected by placing the part in cold water, is usually manifest by pallor, occasionally by cyanosis, rarely by rubor. All colors may be present at the same time.

Permanent, uniform, cyanotic discoloration of the skin distal to the wrists and distal to the ankles usually indicates acrocyanosis. A persistent bluish to bluish-red mottling of the skin of the feet and the legs, known as "livedo reticularis," is particularly noticeable on exposure to cold. Persistent cyanosis of an individual digit is often a manifestation of thrombosis of a digital artery and frequently, but not invariably, precedes gangrene.

A brownish-black discoloration of the legs is a "stasis pigmentation" characteristic of chronic venous insufficiency.

Ulceration, Gangrene

Gangrene caused by chronic occlusive arterial disease usually affects the digits, the first toe being most commonly involved. All digits and the distal parts of the foot, less commonly

the hand, may be affected in extensive gangrene. Gangrene or ulceration of more proximal parts, such as the leg or the heel, may follow trauma.

In Raynaud's disease, small necrotic ulcerations may develop in the tips of the fingers.

Ulceration of chronic venous insufficiency characteristically affects the inner part of the leg just above the ankle, occasionally the outer side of the ankle.

In sudden arterial occlusion, gangrene may be extensive because of severe spasm of the collaterals. However, the more distal parts are usually involved.

An ulceration may develop in any area of the extremity as a result of an arteriovenous fistula and often resembles that due to chronic venous insufficiency. Characteristically, the local skin temperature is elevated.

Swelling and Edema

The following are of circulatory origin:

Deep thrombophlebitis causes acutely developing edema, thrombosed veins are palpable, tender and painful; temperature is elevated slightly.

Chronic Venous Insufficiency. Varicose veins, stasis dermatitis or ulceration, pitting type of edema.

Lymphedema. Gradual progressive extension proximally over a period of weeks, months, or years. If of recent origin, pitting is detected; if extensive and of long duration, the tissues are fibrotic, hypertrophied, and resistant to pitting.

Lipedema is a condition in which diffusion of fluid occurs from the small vessels into surrounding tissues when the latter contains excessive fatty deposits. Lipedema of the legs affects women; appears first during adolescence; the enlargement of the limbs is generalized and symmetric; and frequently its development is associated with a gradual increase in weight. Often a familial tendency is noted, and the adipose tissue is of a peculiar loose texture. The skin and the subcutaneous tissue are soft and pliable The enlargement of the extremity is nonpitting unless edema is extensive and usually at the day's end Generalized obesity may or may not be present

Arteriovenous fistula causes edema because of increased pressure in the veins. Increased oxygen content of the venous blood is diagnostic.

Chronic Occlusive Arterial Disease. A prolonged dependent posture is assumed by the patient to get relief from pain, thereby causing edema. Because the veins in thromboangiitis obliterans are often occluded and effect venous stasis, edema is more common in this condition than in arteriosclerosis obliterans.

Other causes of swelling of the extremities must be differentiated, including systemic (renal, cardiac, hypoproteinemia) and local (tumor) causes.

Temperature

Increased blood flow to the skin is perceived by the examiner's hand as warmth, whereas decreased blood flow is the cause of cold skin The temperature of the skin per se does not indicate the state of circulation in the entire extremity. Many people with abnormally cold skin have normal blood flow in the remainder of the limb. The distal portions of extremities, the digits, show variations in temperature under different conditions, but all digits at any occasion have temperatures which are almost identical. When one extremity displays a skin temperature lower than the opposite extremity, its circulation can be regarded as impaired. By passing the examiner's hand over the extremity from the proximal to the distal end, a sudden change in temperature defines the level of circulatory impairment. One may view with suspicion a single toe whose skin is colder than that of the other toes. When coldness of the skin occurs symmetrically in both extremities, the circulation can be implicated only in the light of other findings, and if diminished temperatures are demonstrated repeatedly under basal conditions. Cold skin is associated with chronic occlusive arterial disease and the pallor phase of Raynaud's disease. A warm skin accompanies erythermalgia

The subjective sensation of "warmth or burning" is a paresthesia occurring frequently in patients of advanced age and does not necessarily indicate a circulatory disturbance. When associated with a warm skin, erythermalgia must be considered.

Arterial Pulsations

The pulsations can be felt in the upper extremity in the subclavian, the axillary, the

brachial, the radial and the ulnar arteries; in the lower extremity in the femoral, the popliteal, the dorsalis pedis and the posterior tibial arteries. The degree of expansile force is estimated and recorded. Occasionally, arterial pulsations may be detected in an abnormal situation, particularly about the wrist, the knee and the ankle, and may signify the extent of collateral circulation.

Allen's Test. This is used to determine arterial occlusion distal to the wrist and the ankle when pulsations at these joints are present. When one artery is obstructed, postural color changes, and lowered skin temperature may be absent, owing to free arterial inflow through the uninvolved vessel.

The test is performed as follows: Both hands of the patient are elevated in front of the examiner. If the ulnar artery is suspected, each radial artery is compressed at the wrist. The patient squeezes the blood out of the wrist by clenching the hands as tightly as possible for about 10 seconds, then opens them. The return of color is rapid in the unaffected hand. If the ulnar artery is occluded, pallor is persistent for some time or until the radial artery is released.

The radial, the dorsalis pedis and the posterior tibial arteries may be similarly tested. In examining the feet, elevation and dependency are substituted for clenching and opening the hands, respectively.

Varicose Veins

Varicose veins are venous channels which have become large and distended by abnor-

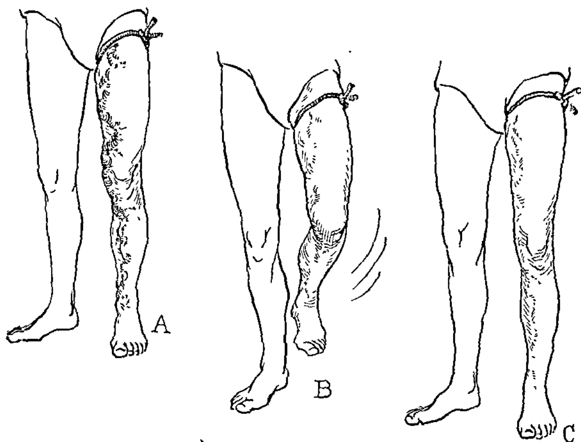


Fig. 251. Perthes' test for incompetence of superficial and deep veins. A tourniquet about the thigh compresses the long saphenous vein, preventing reverse flow past this constriction. The patient then exercises this extremity briskly. Normally, blood flow in the venous system is aided by muscular action so that it goes from the superficial to the deep system. If the superficial varices disappear when the leg is exercised, the valves of the communicating veins are competent, and only the saphenous valves are incompetent. If the varices do *not* disappear with exercise, the valves in the communicating veins as well as those in the saphenous vein are incompetent. If the veins become more prominent with exercise and pain appears, the deep veins are obstructed, and ligation of the superficial system is contraindicated.

mally and chronically increased intraluminal pressure resulting from (1) *incompetent venous valves* permitting gravitational back flow of blood; (2) *obstruction to blood flow* (e.g., thrombophlebitis, tumor compression and formation of collaterals; and (3) *arteriovenous fistula,* causing the blood to flow directly from the artery into the vein.

Superficial thrombosed veins can be palpated as thickened cords. When thrombophlebitis is acute, a narrow zone of redness overlies the tender palpable vein. Deeply situated veins cannot be palpated, but their patency may be determined by tests of filling and emptying applied to superficial veins.

A vein normally fills within 10 seconds when it is brought to a dependent position after elevation. Any delay in filling, assuming that the venous valves are competent, is presumptive evidence of impaired arterial circulation. A vein normally empties when it is elevated a few centimeters above the level of the heart. Failure to empty signifies abnormally increased intravenous pressure.

Tests to Determine Incompetency of the Saphenous and the Communicating Veins and Occlusion of the Deep Veins. These tests must be performed together. Before removing or obliterating the superficial channels, it is important to establish that deep channels are adequate.

PERTHES' TEST. While the patient is standing, a tourniquet is applied about the upper thigh sufficient to compress only the long saphenous vein. Then the patient walks about briskly, and the prominence of the varicose veins is noted. Normally, muscular action should empty blood from the superficial system through the communicating veins into the deep system. Therefore, disappearance of varices indicates that valves in the communicating veins are competent, and varicosities are due solely to incompetent saphenous valves. If varicosities do not disappear while walking both saphenous and communicating valves are incompetent. If the varicosities become distended and prominent, and the patient experiences pain while walking, the deep veins are obstructed, and the valves of the communicating valves are incompetent. This may also indicate incompetency of communicating veins between the long and the short saphenous veins.

THE PRATT TEST. This determines the location of incompetent communicating branches.[2] The recumbent patient elevates the leg and empties the veins. A tourniquet compresses the long saphenous vein at the upper thigh. An elastic bandage is applied from the toes to the tourniquet. The patient stands erect, and the bandage is slowly unwound from above downward. Reflux blood from above is prevented by the tourniquet, so that the appearance of a bulge or a blowout indicates the site of an incompetent communicating vein. This is marked with an indelible pencil. A second bandage is applied from the level of the tourniquet down to and compressing the bulging vein. Then the first bandage is again unwound downward to the next blowout, which is again marked and compressed by the second bandage. This procedure is continued until all blowouts are identified. Removal of all incompetent communicating veins is necessary to prevent recurrences. While the bandages are applied, severe pain and swelling in the calf indicate occlusion of the deep veins.

SPECIAL TESTS OF PERIPHERAL CIRCULATION

Many procedures have been devised with a view toward defining accurately the condition of vessels of the extremities. Only a few of these tests are of clinical value and are used mainly as adjunctive to a direct examination.

ANGIOGRAPHY

Arteriography. Injection of a radiopaque substance into the lumen of an artery will permit roentgenographic visualization of the component parts of this artery. The substances commonly used are:

Skiodan. Because large amounts (50 cc.) can be injected safely, it is preferable for visualization of large vessels. General anesthesia is required, because arterial spasm, pain and muscle spasms are caused by the drug. Hypertension is a contraindication.

Diodrast. Although it produces no pain or arterial spasm, its usefulness is limited by the smaller maximal dose (20 cc.).

Thorotrast (Stabilized Thorium Dioxide Sol). This substance is painless and provides

[2] Pratt, G. H.: Test for incompetent communicating branches in the surgical treatment of varicose veins, J.A.M.A. 117:100, 1941.

excellent visualization of arteries of the upper extremities by injection of only 5 to 12 cc. The possibility of delayed untoward effects due to radioactivity should be borne in mind.

TECHNIC FOR THE UPPER EXTREMITIES. The patient lies recumbent upon the x-ray table, the extremity lying outstretched and supinated upon a film holder. A blood pressure cuff is placed about the upper arm. Under local anesthesia, the needle of the syringe, containing the radiopaque material, is inserted into the lumen of the brachial artery. The blood pressure cuff is inflated above the systolic pressure, the radiopaque material is injected, the needle is withdrawn quickly, and the first film is exposed immediately. Next, the cuff is deflated for a few seconds to the level of the diastolic pressure and reinflated. This permits the radiopaque material to be carried more distally. Another roentgenogram is made. The procedure is repeated several times until adequate visualization of the smallest peripherally situated vessels and the veins has been obtained.

TECHNIC FOR THE LOWER EXTREMITIES. The femoral artery is palpated and marked below the inguinal ligament Under local anesthesia, it is punctured, occluded above the point of puncture by digital compression, the drug is injected, and the x-ray film is exposed. The compression is released for a few seconds, again the artery is occluded, and another film is made. The procedure is repeated several times.

When an arteriovenous fistula is present, the contrast medium is carried rapidly into the venous circulation If this condition is suspected, it is advisable to maintain compression of the proximal portion of the vessel until at least 3 films have been exposed.

INTERPRETATION OF ARTERIOGRAMS OF THE EXTREMITIES. A normal arteriogram is characterized by (1) a smooth, uninterrupted contour of the lumens, (2) a direct course of the vessels and (3) only a minimum of collateral vessels. A vessel in certain situations normally changes its course, but the change of direction is gradual, not abrupt. Spasm of an artery is characterized by smooth diminution in caliber as the point of occlusion is approached, then a gradual resumption of caliber beyond the constriction. The appearance typically varies from film to film.

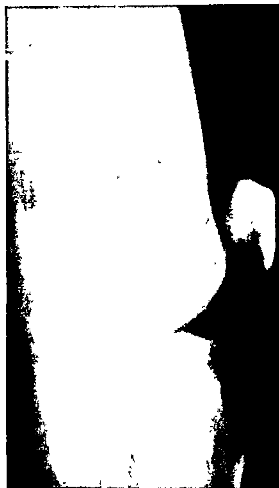


FIG. 252. Aneurysm of popliteal artery demonstrated by angiography.

Collateral arteries become prominent in number and size as main arteries are occluded. These compensatory vessels are identified by (1) an irregular twisting and turning course, (2) variation in size in the same locale, (3) their purposeless crossing and recrossing, (4) a transverse course in areas where they are profuse, and (5) anastomoses. When an artery is partially occluded, an anastomotic branch often forms proximal to the point of occlusion, can be seen running alongside the parent artery, and re-enters the artery distally. When a digital artery is occluded, numerous minute collateral arteries pass laterally from the companion artery to the diseased artery. When radial and ulnar arteries are occluded, the interosseous artery, which ordinarily is never visualized beyond the level of the wrist, extends distalward to compensate for loss of blood supply. Complete occlusion of a large artery throughout its course may be compen-

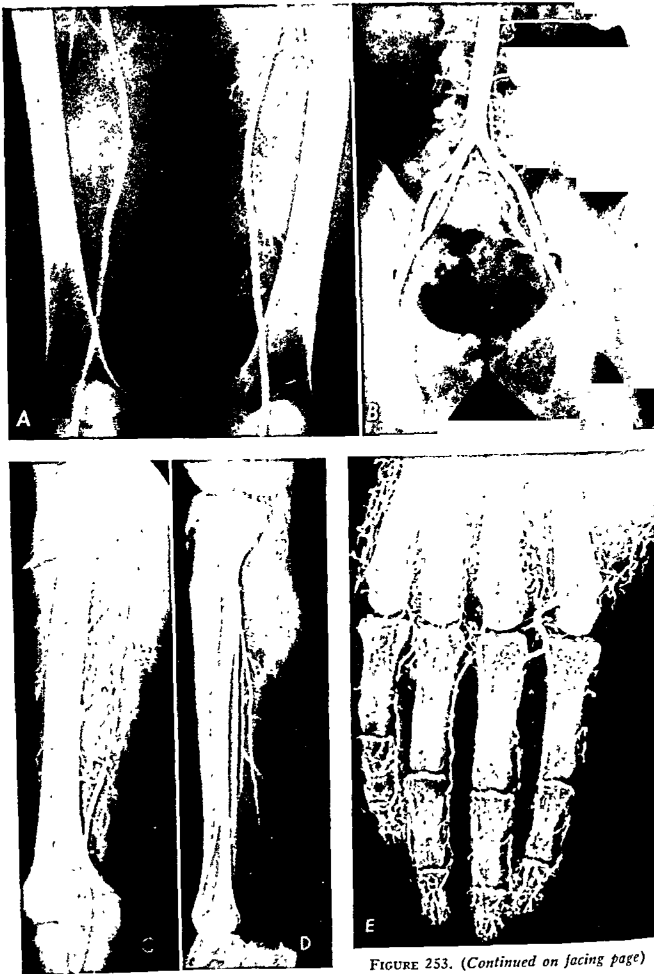


FIGURE 253. (*Continued on facing page*)







FIG. 253. Arteriograms, their characteristics in disease. (A and B) The normal arteriogram. The aorta, the iliac and the femoral vessels are of uniform caliber and are smooth and regular in outline (C and D) Arteriosclerosis obliterans, showing complete occlusion of the mid-portion of the superficial femoral artery. There is good distal filling below the obstruction but all of the [illegible] their [illegible] rrom- [illegible] e has [illegible] This arteriogram of an affected hand shows occlusion of most of the digital vessels with collateral branching which is nature's way of maintaining adequate circulation. (F and G) Acute arterial occlusion of the popliteal artery which followed trauma. Except for the occluded segment, the vessels are fairly normal in appearance. (H and I) Arteriosclerotic aneurysm of the popliteal artery. The arteriogram demonstrates the channel through the aneurysm. Clinically, the aneurysm is much larger since most of its interior is filled with clot. (J) Traumatic arteriovenous fistula: This is an abnormal communication between the superficial femoral vessels due to a gunshot wound. Note the enormous dilatation of the proximal vein. (Dr. John C. Ivins, Mayo Clinic, Rochester, Minn.)

sated by formation of numerous large branches arising above the site of occlusion.

Arteriographic Characteristics of Peripheral Vascular Disease

THROMBOANGIITIS OBLITERANS. Patchy distribution of changes chiefly affecting the peripherally situated small caliber vessels is characteristic. Adjacent vessels may be affected to a different degree. Involvement is revealed by a moth-eaten, irregular filling of defects, narrowing of the lumen, irregular changes in caliber from segment to segment, and an irregular, rapidly changing course. When occlusion is complete, the point of obstruction is rounded rather than abrupt. Collateral vessels are numerous, more so than in arteriosclerosis.

ARTERIOSCLEROSIS. Typically, the roentgenographic features are: extremely irregular lumens, narrow caliber, moth-eaten contour, and a moderate number of collateral vessels.

ANEURYSM The sac often is filled with clotted blood, and the radiopaque material fails to enter the cavity. Nevertheless, the shadow of the vessel is interrupted abruptly at the level of a rounded soft tissue density, and the distal vessel is frequently visualized beyond the mass. Occasionally, a thin line of increased density forms a border about the sac.

ARTERIOVENOUS FISTULA. The arteriographic features are: increased size and tortuosity of arteries leading to the fistula, pooling of the radiopaque substance in the region of the fistula, absence of filling of the arteries distal to the fistula, and visualization of the veins more rapidly than usual.

RAYNAUD'S DISEASE. The distal portions of the digital arteries fail to fill, and their caliber is diminished.

BONE TUMORS. In benign tumors, the blood vessels may be displaced but are of normal size and number. Malignant tumors often exhibit numerous vascular pedicles and a profuse network of newly formed vessels

ARTERIAL OCCLUSION The site of obstruction can be localized prior to surgical intervention. Where no surgery is contemplated, arteriography is contraindicated because of the possibility of provoking spasm of the collaterals

Venography. The veins may be visualized by the *indirect method* (injection into the artery) or the *direct method* (injection into the vein). Venography has very limited clinical use. For example, one may identify a point of obstruction in the subclavian vein as the cause of swelling of the upper extremity; many tortuous collateral veins are visualized. The main usage of venography is in the study of anatomy and direction and rapidity of blood flow. The method has been advocated for the diagnosis of acute venous thrombosis of veins of the lower extremity.[3] The diagnostic features are: (1) incomplete filling of the veins, (2) dilatation of the superficial veins, indicating thrombosis of the deep veins, and (3) increased resistance to injection.

OSCILLOMETRY

The oscillometer is a device for measuring the amplitude of pulsation in the extremities. It consists of a blood pressure cuff connected by tubing with an instrument containing an oscillating needle. The cuff is wrapped about the extremity and is inflated. By adjusting the valves the needle moves over a scale with each pulsation. The extent or the number of marked units over which the needle passes depends upon not only the volume of pulsation but also the amount of pressure within the cuff. Therefore, the cuff is inflated above the value of the systolic blood pressure and is deflated at levels of 10 mm. of mercury, the oscillation of the needle being recorded at each level. The maximum number of units of needle excursion found at any level constitutes the *oscillometric index*.

The oscillometric index varies greatly between individuals. It may vary somewhat in the same individual at different sittings. It supposedly is useful for quantitatively measuring the progress of arterial disease in the extremities. However, it does not indicate the state of the collateral circulation or the nutrition of the part. Skin temperature determinations more accurately determine changes in the volume of blood flow to the extremities, particularly after sympathectomy.

Normal oscillometric readings are:[4] arm 4 to 20, elbow 2 to 12, wrist 1 to 10, palm 0 to 2,

[3] Welch, E. E, Faxon, H. H, and McGahey, C. E: The application of phlebography to the therapy of thrombosis and embolism, Surgery 12.163, 1942.

[4] Samuels, S. S.: The value of oscillometry; in the study of circulatory disturbances of the extremities, J.A.M.A. 88:1780, 1927.

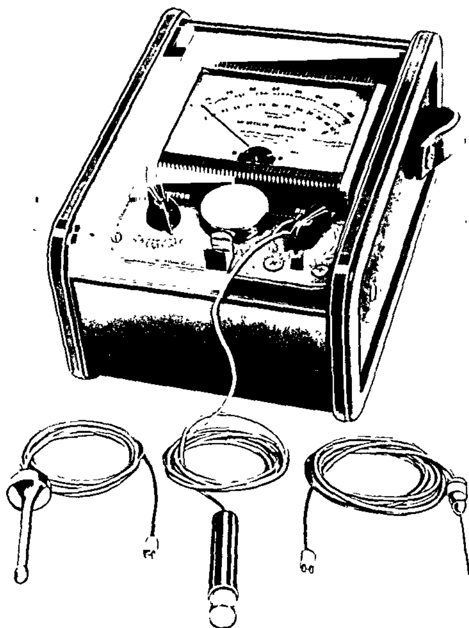


Fig. 254. Portable skin thermometer. (McKesson Dermalor, McKesson Appliance Co., Toledo, Ohio)

thigh 4 to 16, upper leg 3 to 12, ankle 1 to 10, and foot 0.5 to 2.

Skin Temperature Determinations

The temperature of the skin indirectly reflects the condition of the circulation of the extremities. It is an index to the amount of blood flow to the skin. The instrument used consists of two thermocouples. One thermocouple is maintained at a constant temperature. When the temperature of the other differs from that of the first, a current flows over the wire connecting the thermocouples This causes deflection of the string galvanometer which reflects a light beam on a graduated scale. The latter indicates the degrees of temperature of the surface upon which the thermocouple is placed.

The normal surface temperature of feet varies from 24° to 35° C., being slightly lower in patients who have cold feet without circulatory impairment. Under standard environmental conditions (77° F. or 25° C. and 40% humidity), the temperature of the toes is slightly above room temperature. Temperature of the fingers is between 32° and 35° C. The following facts are of importance in interpreting the results:

1. A difference in temperature between two symmetric parts exceeding 2° C. indicates impaired circulation. This applies also to differences between digits of one foot or hand.

2. A sharp change in temperature of the skin under basal conditions indicates a change in circulation to the skin. (Examples: sympathectomy causes abrupt rise in temperature; arterial occlusion causes sudden drop.)

3. When the patient is moved from an environment of basal conditions to a temperature at or beyond 30° C., maximal vasodilatation normally causes the skin temperature of fingers to rise substantially beyond that of room temperature. Failure to obtain this effect indicates defective arterial circulation.

4 Induction of artificial fever (typhoid vaccine intravenously) raises skin temperature of the digits to or beyond 34° C. Failure to do so indicates impaired arterial circulation.

Reflex Vasodilatation

When one extremity is warmed, for example by immersing it in warm water, vasodilatation is induced in the digits of the other extremities as evidenced by a rise in skin temperature. This indirect vasodilatation depends upon the return of blood from the warmed extremity to the general circulation and on the integrity of the sympathetics. Failure to obtain an increase in temperature of the skin of the fingers indicates defective arterial supply in which the vasoconstrictive component is negligible. An abrupt rise in skin temperature suggests a vasoconstrictive factor which can be overcome by sympathectomy.

Histamine Flare Test

When histamine is injected intradermally, a wheal surrounded by a zone of erythema and accompanied by itching develops within 10 minutes. In the presence of impaired arterial circulation development of the wheal and the flare is delayed or absent. The test is often used in the lower extremity to investigate the adequacy of circulation at various levels and to determine the site of amputation. For example, a drop of a 1:1,000 solution of histamine acid phosphate is injected intradermally over the dorsum of the foot, 6 inches below the knee, at the lower third of the thigh, and at the midthigh. Formation of a wheal indicates that sufficient collateral circulation for healing has been established.

Anesthesia

General anesthesia causes widespread vasodilatation. Spinal anesthesia will effect vasodilatation only in the area of sensory loss. Likewise, injection of procaine into a peripheral nerve will cause a rise of surface temperature in the anesthetic zone. For practical purposes, a sympathetic block determines the maximal vasodilatation of the part. Normally, the lower limit of surface temperature of the great toe after sympathetic block should be approximately 31.5° C. When this reading is lower or a minimal rise of surface temperature occurs after a sympathetic block, organic arterial occlusive disease is the main cause of impaired circulation. An abrupt rise in skin temperature implicates arterial or arteriolar spasm as the offending factor, which can be overcome by sympathectomy.

ARTERIOSCLEROSIS OBLITERANS

Arteriosclerosis obliterans is characterized by occlusion of large and medium-sized arteries, mainly in the lower extremities, by extensive atheromatous formations and thrombi causing peripheral ischemia.

Etiology

The cause of arteriosclerosis is unknown. Theories include:

1. **Mechanical cause**, a result of continual pulsating trauma of the arteries throughout the years of life. This explains why hypertension is more frequently and earlier associated with arteriosclerosis; atheromas are commonly found around the orifices of branches of the aorta; and the more severe lesions are situated in the lower extremities where blood pressure is higher.

2. **Metabolic disturbance**, particularly of cholesterol and lipoids. It explains why lipoids are contained in large amounts in atheromas and correspond in proportions of different constituents to those found in the blood plasma; the lesion may be reproduced in experimental animals on a diet of cholesterol or animal fat; disorders manifested by lipemia are frequently complicated by severe and early atherosclerosis. *Diabetes mellitus* is a predisposing factor to severe and premature arteriosclerosis obliterans.

Pathology[5]

The lesions of arteriosclerosis obliterans are

[5] Allen, E. V., Barker, N. W., and Hines, E. A., Jr.: Peripheral Vascular Diseases, Philadelphia, Saunders, 1948.

situated predominantly in the large arteries of the lower extremities, such as the iliac, the femoral and the popliteal, and in medium-sized arteries such as the anterior tibial and the posterior tibial, but rarely are the small arteries, such as the digital and the plantar, involved.

Grossly, the arteries are enlarged, irregular, tortuous and rigid. Sectioning reveals a thin medial coat, calcium deposits in the media or at the base of atheromas, irregular atheromatous formations extending a considerable distance into the lumen, and partial or complete occlusion of the lumen by gray or red thrombi.

The atheroma is an elevated yellow plaque which consists of connective tissue thickening of the intima and phagocytic cells loaded with fat. These yellow patches tend to develop on one side of the artery and become confluent. As the atheroma enlarges, microscopically one sees large acellular, hyalinelike areas, irregular deposits of fat, fatty acids and cholesterol, lipophages, fibroblasts and fibrous tissue. Often a deposit of calcium is situated at the base of the atheroma. The surface is eroded and forms an irregular ulcer upon which a mural thrombus is deposited. Succeeding layers of thrombi are laid down and gradually occlude the artery. The deeper layers of thrombus become organized and merge imperceptibly with the atheroma. Recanalization occurs in some thrombi.

Other pathologic features, the result of ischemia, include atrophy and thinning of the skin, muscle atrophy with fibro-fatty replacement, loss of subcutaneous fat, ulceration and gangrene Ischemic neuritis is usually associated with extensive arterial occlusion of larger arteries.

Pathogenesis

The thrombosis will occur if the atheroma is large and its surface rough and ulcerated. It is more likely to develop in the presence of a blood dyscrasia, particularly polycythemia vera An increased susceptibility to thrombosis may develop after operations, injuries and infectious diseases, especially in cases of early atherosclerosis.

Pathologic Physiology

The obstructive lesions involve large arterial trunks predominantly. The degree of ischemia is proportionate to (1) the extent of occlusion, including obstruction of potential collateral channels, (2) the height or most proximal point of the occluding process, and (3) the rapidity of developing occlusion. Because the arterial tree is rigid, and although extensive collateral channels can develop, the potential for formation of collaterals is not as great as in thromboangiitis obliterans.

Arteriolar constriction can be augmented by cold and tobacco; heat effects vasodilatation. The degree of arteriolar dilatation may be surprisingly great because the small arteries are not much affected by arteriosclerosis.

Arterial pressure is lowered beyond the point of obstruction, as evidenced by lower skin temperature and by the elevation-dependency test. On elevation, the extremity becomes abnormally pale; on lowering it, there is a delay in the return of color to the skin. Continuing the dependent position, there appears a rubor, the result of capillary atony. The capillaries are atonic because of ischemic malnutrition. Edema of the foot develops not only because of this atony but also because the patient constantly holds the foot in the dependent position to relieve pain.

The presently accepted theory of mechanism of production of intermittent claudication is as follows: Because of inadequate blood flow through muscles during exercise, a pain factor, "factor P," accumulates and stimulates the sensory nerve endings. The pain disappears when collateral supply becomes adequate, or when the affected muscle atrophies and becomes fibrotic.

Clinical Picture

The symptoms are those due to ischemia, and the findings are those of occlusive arterial disease. They are almost invariably confined to the lower extremity, where they may come on gradually or abruptly.

Symptoms

1. *Intermittent Claudication.* This is the earliest symptom when arterial occlusion develops slowly. It is usually unilateral at first but eventually becomes bilateral. The extent of arterial occlusion determines the rapidity with which pain appears while walking. The pain is usually situated in the foot or the calf, occasionally in the thigh, the hip and the lower back.

2. *Rest Pain.* In severe degrees of occlusion.

the ischemia is made worse by the horizontal position. A severe, aching, persistent pain is noted in the toes, the foot, or the leg, especially at night. The patient sits up, rubs his foot and holds it dependent to relieve the pain. An acute arterial occlusion will cause rest pain. Because this type of pain frequently is the forerunner of ulceration and gangrene, it is often called "pretrophic pain."

3. *Pain of ulceration and gangrene* is a persistent moderate or severe pain similar to rest pain but usually confined to the region of ulceration or gangrene.

4 *Pain of ischemic neuritis* is a paroxysmal, severe lancinating pain which extends over the entire extremity and often follows the distribution of the peripheral nerve. Between paroxysms, a steady ache or burning sensation may be noted. This type of pain occurs commonly in the diabetic, when the upper femoral or the external iliac arteries are occluded, or after a sudden severe arterial occlusion.

5. *Paresthesias and anesthesia* are common and probably are due to ischemic neuritis. A complete stocking type of anesthesia of the toes or the foot or the entire leg develops after extensive acute arterial occlusion. It is a grave prognostic sign which indicates that gangrene will develop.

6 *Cold Sensitivity.* The patient complains of excessive coldness, even with slightly lowered environmental temperature.

Findings

1. *Impaired arterial pulsation* is determined by palpation. The dorsalis pedis is normally absent in 8 per cent of subjects and of itself is not necessarily significant. Regardless of the degree of atherosclerosis, reduction or absence of arterial pulsations is necessary to the diagnosis of arterial occlusion.

2. *Color Changes.* The toes may be red bluish, or pale. Following recent acute occlusion, the foot may be extremely pale. Changes of color denote severe degrees of arterial occlusion.

3. *Postural Color Changes.* Abnormal pallor on elevation and delay of return of color followed by rubor on dependency, associated with delay in venous filling is pathognomonic of occlusive arterial disease.

4. *Temperature Changes.* Lowered skin temperature, especially if a difference between the two feet is noted, is significant.

5. *Trophic Changes, Ulceration, Gangrene and Infection.* The toes are scarred and shrunken. Ulceration and gangrene are often the result of avoidable trauma. When spontaneous, ulceration and gangrene usually develop in the terminal portions of the toes about the nails but may extend to involve the entire toe, occasionally the foot, rarely the leg. Extensive gangrene denotes an acute or extensive occlusion which implies insufficient collaterals.

Ordinarily, ulceration and gangrene are of the dry type without systemic reaction. In diabetics, however, the lesions are often moist and infected, with rapid spread through the lymphatics and septicemia.

Minor infected lesions, such as paronychias, occur frequently. They may heal, or they may develop into gangrenous or ulcerative lesions, particularly if subjected to surgical interference.

6. *Atrophy of muscles, skin and other soft tissues*

7. *Osteoporosis*

8. *Edema* of the foot and the leg

Laboratory Findings

Roentgenologic Findings. Calcification of the arteries is visualized. One type is the diffuse regularly distributed type of Mönckeberg calcification (in the medial coat). The second type is the localized patchy dense deposits of calcium usually located at the base of large atheromata. It more often is found in the midportion of the femoral artery, a common site for initial arterial occlusion.

Diabetes mellitus studies include urinalysis, fasting blood sugar and the glucose tolerance test.

Determination of Plasma Lipoids. Where differentiation from thromboangiitis obliterans in the middle-aged patient is difficult, elevated values for plasma lipoids may favor a diagnosis of arteriosclerosis obliterans The normal upper limits for these lipoids per 100 cc. of plasma are: cholesterol, 250 mg., cholesterol esters, 175 mg., total phospholipids, 275 mg., total fatty acids, 400 mg., and total lipoids, 650 mg. Values exceeding these signify lipemia

Skin Temperature. Elevation of skin temperature in response to sympathetic block will indicate the benefit to be derived from sympathectomy.

Oscillometry has been used to determine the degree of arterial pulsation. These readings are unreliable, especially because the cuff cannot be applied at the same level each time. Digital palpation is more reliable.

Histamine wheal test is used to determine circulatory adequacy at various levels.

Arteriography will localize the point of occlusion but is not advisable, because irritation by the radiopaque substance may cause damage to collaterals.

Diagnosis

The features of occlusive arterial disease are absent pulsations, postural color changes, intermittent claudication and possibly digital ulcers or gangrene. The patient often is above 50 years of age, both sexes are affected, calcification of large arteries is often visible in roentgenograms, and plasma lipoids may be elevated. Diabetes mellitus is strong evidence in favor of arteriosclerosis obliterans.

Differential Diagnosis. This topic is covered under the subject of Thromboangiitis Obliterans.

Prognosis

In this progressive, degenerative vascular disease, the outlook for survival of the extremity is not good. However, if arterial occlusions are not too extensive or too frequent, it is possible for adequate collateral circulation to develop and compensate for loss of the main artery. Once gangrene develops, the chances for saving the extremity is poor, especially when diabetes is associated. The life expectancy is shortened; many die of coronary occlusion. The mortality rate following amputation varies from 4 to 20 per cent, the rate being lowered by higher levels of amputation.

Treatment

Prophylaxis is mainly the concern of the internist. In view of results of experimental work to date, a diet low in fat and cholesterol is theoretically valuable in delaying progression of atherosclerosis. Because a low fat diet is deficient in Vitamin A, the latter should be supplied.

A small amount of thyroid is given to those patients with a low basal metabolic rate.

The orthopaedic surgeon in recent years, particularly since World War II, has been delegated more responsibility in the handling of occlusive arterial disease. Management naturally falls within the province of the man who ultimately may rehabilitate the patient by amputation and fitting and use of a prosthesis.

The orthopaedic management is described in the next section on Thromboangiitis Obliterans.

THROMBOANGIITIS OBLITERANS
(Buerger's Disease[6, 7])

Thromboangiitis obliterans is a disease of unknown origin, occurring predominantly in young and middle-aged males, often heavy smokers. It is characterized pathologically by recurring episodes of inflammation and thrombosis in medium-sized arteries, principally in the lower extremities, and clinically by symptoms and findings of sudden or chronically progressive arterial occlusion. The condition frequently comes to the attention of the orthopaedic surgeon in attempting to define the cause of pain in the lower extremities. Recognition at an early stage is of importance in halting progression of the condition and preventing its serious complications.

Etiology

Although the exciting cause is unknown, the following are predisposing factors:

Age. Mainly in young or middle-aged adults between 25 and 45 years of age

Sex. Males almost exclusively. Experimental work has shown that estrogenic hormone will prevent ergotamine induced gangrene in rats.[8]

Race. All races, Jewish race seems to be predisposed.

Tobacco. The majority of patients are heavy smokers. The disease progresses if smoking is continued; if smoking is discontinued, new vascular occlusions are rare. Nicotine causes vasoconstriction and occlusion.[9]

[6] Buerger, L · The Circulatory Disturbances of the Extremities, Philadelphia, Saunders, 1924.

[7] von Winiwarter, Felix· Ueber eine eigenthumliche form von endarteritis und endophlebitis mit gangran des fusses, Arch. f. Chir 23 202, 1879

[8] McGrath, E J. C Experimental peripheral gangrene; effect of estrogenic substance and its relation to thrombo-angiitis obliterans, Arch. Int. Med 55. 942, 1935

[9] Winroth, L. A, and Herzstein, J.: Relation of tobacco smoking to arteriosclerosis obliterans in diabetes mellitus, J.A.M.A. 131:205, 1940.

Weather. The condition is worse in winter months when cold increases vasoconstriction.

Blood Changes. An increased tendency to coagulation is found in many patients.

Infection. Various investigators have recovered organisms which, when injected into experimental animals, occasionally produce intimal proliferation and thrombosis. Removal of foci of infection often will reduce pain of claudication

Pathology

Medium-sized or small arteries, especially of the lower extremities, are involved. The large arteries are affected late and only in the severely progressive case Less often the small and the medium-sized veins are affected. The lesions are segmental and are sharply demarcated from intervening normal vessel. At the site of pathology artery, vein and nerve are often bound together by fibrous tissue. Microscopically, an inflammatory panarteritis or panphlebitis with thrombosis is seen. The adventitia displays aggregations of lymphocytes about the vasa vasorum and fibrous hyperplasia The muscle layer is atrophic The intima is thickened by endothelial proliferation. The lumen is filled with a thrombus which varies from a fresh clot to one that is completely organized. Openings in the occluding mass indicate an attempt at recanalization Extensive collaterals develop about the lesion.

This lesion is distinguished from periarteritis nodosa by the greater extent of the lesion, the absence of necrosis in the media, the absence of aneurysms, and the invariable presence of an occluding mass

Clinical Picture

Symptoms and findings involve the lower extremities in 95 per cent of cases.

Symptoms

These include pain, a sensation of coldness, and abnormal sensitivity to cold temperatures.

1. **Pain.** Various types of pain are experienced as follows:

A. Intermittent Claudication. This is the most common and often the earliest complaint. It usually is produced by exercise, relieved by rest, is accentuated by cold and always occurs distal to the point of occlusion, especially in the arch of the foot and the calf of the leg.

B. Rest Pain. This consists of a severe, gnawing ache in the toes, usually appearing after an acute occlusion and remaining severe for days or weeks. It is worse at night, aggravated by elevation and reduced by dependency (the reverse may occur). It is caused by ischemia of tissues, including the sensory nerve terminals. Because it is often a prelude to ulceration or gangrene, frequently it is termed "pretrophic pain."

C. Pain of Ulceration or Gangrene. The pain often becomes intense and persistent

D. Ischemic Neuritic Pain. This often occurs late in the course after involvement becomes extensive and large arteries are occluded It is a paroxysmal severe pain associated with various paresthesias and is widespread through the extremity. It resembles lightning pains. During paroxysms blanching, cyanosis, or mottling of the skin and excessive sweating are due to sympathetic stimulation Between paroxysms, the pain may persist as a dull ache.

E. Thrombophlebitic Pain. This is usually mild and aggravated by use of the limb or pressure on the vein. It disappears with subsidence of the inflammation.

2. **Coldness, Sensitivity to Cold.** The toes and the foot are subjectively cold and numb. These symptoms are accentuated by slightly cold temperatures, which at the same time cause blanching and cyanosis.

Findings

Objective findings pertain to identifiable arterial occlusion and the effects of circulatory insufficiency.

1. **Impaired Arterial Pulsation.** The dorsalis pedis and the posterior tibial pulsations are reduced or absent in a majority of patients One must remember that the dorsalis pedis is absent normally in 8 per cent of people, and its absence alone does not necessarily imply occlusive disease.

Absence of popliteal and femoral pulsations is less frequent. Radial or ulnar pulsations are absent in 40 per cent of patients.

Occlusion can occur distal to the ankle or the wrist beyond the point of palpable pulsations This can be determined as follows, inquiring into the patency of the ulnar artery as an example: The radial artery is lightly compressed. The patient clenches the fist, squeezing out the blood, then opens the fist. Normal

color should return immediately. In ulnar occlusion, pallor persists. The radial, the dorsalis pedis and the posterior tibial arteries can be tested similarly.

2. **Color Changes.** With elevation, the part is abnormally blanched. With dependency, after a delay of 5 to 30 seconds, color returns, and an abnormal rubor appears (R-P sign). The postural color changes are typically asymmetric.

3. **Temperature Changes.** Asymmetric coldness is palpable. The temperature may vary from toe to toe.

4. **Ulceration and Gangrene.** This may develop spontaneously or may follow mechanical, chemical or thermal trauma. A small ulceration commonly appears about nail margins. Gangrene is commonly dry and involves one digit, less often the foot. Cellulitis and spreading infection are uncommon.

5. **Edema** may be caused by thrombophlebitis and ischemic necrosis of small distal vessels.

6. **Trophic Changes.** Nails are thick and deformed, and digits are shrunken. Thin atrophic skin is more common in arteriosclerosis. Bony structures are osteoporotic.

7. **Superficial Thrombophlebitis.** Occurs in 40 per cent of patients. It lasts from 1 to 3 weeks. Thrombophlebitis when seen in association with occlusive arterial disease definitely identifies the latter as thromboangiitis obliterans.

Special Tests

Various procedures are used only occasionally as adjunctive to diagnosis by clinical examination.

1. **Arteriography.** By this means the point of occlusion can be localized. However, the procedure might be condemned on the grounds that the injected solution may irritate and cause spasm of collateral vessels.

2 **Skin Temperature Determinations.** Measurement is done in a room of constant temperature, between 75.2° to 78.8° F. Differences of more than 2° C. in the temperature of various digits are suggestive of local arterial obstruction.

3. **Test for Vasodilatation.** An abnormal degree of arteriolar spasm is present early in the course of the disease. Sympathetic block is the most certain method of effecting vasodilatation in the extremity. A rise in temperature of the part to 86° F. indicates a good response and very little organic obstruction. Lesser degrees of rise correspond to the severity of obstruction and a lesser amount of improvement to be expected from sympathectomy.

4. **Oscillometry** determines the impairment of arterial pulsation and may be used to evaluate the result of treatment.

5. **Claudication time** also determines the effect of treatment. The patient is observed walking at a rate of 120 steps per minute, and the time of appearance of claudication is noted.

Course

Episodes of new occlusions, alternating with periods of remissions take place. Between attacks, collaterals develop. There is a tendency toward improvement and, in most cases, the disease ultimately becomes inactive. Even after gangrene, return to normal activity is possible. The course varies from mild and nonprogressive to severe and slowly progressive.

Diagnosis

Early diagnosis may prevent gangrene. Significant signs include intermittent claudication, superficial thrombophlebitis, abnormally cold extremities with asymmetric color changes, occurrence in a male who is usually a heavy smoker, and evidence of occlusion.

Prognosis

The outlook as to life is good. In regard to loss of limb, the degree is variable but is influenced favorably by discontinuing smoking.

TREATMENT OF CHRONIC OCCLUSIVE ARTERIAL DISEASE

Recommended Measures

The aim is to prevent progression while collaterals develop. The following measures are recommended:

1. **Rest in bed** reduces oxygen need and permits healing of ischemic neuritis, ulcers and gangrene.

2. **Warm environment** encourages vasodilatation.

3. **Smoking should be discontinued permanently.**

4. **Protection against injury**, avoiding constricting clothing, tight shoes, etc.

5. **Avoid extremes of temperature.**

6. **Exercise within limits,** especially in buoyant water or in a whirlpool, will encourage blood flow and reduce thrombotic episodes.

7. **Fever therapy** produces maximal vasodilatation, stops progress of the disease, relieves pain and controls thrombophlebitis. Typhoid vaccine or other foreign protein is given in gradually increasing doses every other day until a temperature of about 102° F. is attained without causing a chill. A series of 8 to 12 injections constitutes a course, and several courses may be required.

8. **Vasodilator drugs** include whiskey and papaverine. The various preganglionic sympathicolytic drugs, such as tetraethylammonium chloride, in effective doses may cause dangerous hypotensive levels.

9. **Reflex heat** by placing the hands in hot water or an electric pad upon the abdomen will reflexly effect vasodilatation in the lower extremities.

10. **Postural exercises** have a questionable effect. The extremity is elevated until pallor appears; next, it is kept horizontal for about 30 seconds; finally, it is held dependent until rubor appears. The exercise is repeated a number of times at each of several sessions per day.

11. **Constant elevation must be avoided** as it may cause dangerous ischemia.

12. **Sander's oscillating bed** by its rocking motion gives postural exercises without effort by the patient.

13. **Sympathetic blocking** with a local anesthetic in oil or in solution by continuous drip through a polythene tube will relieve vasospasm and ischemic pain and may prevent gangrene. Permanent blocking can be secured by injecting 6 per cent phenol.

14. **Sympathectomy** is recommended in all cases. It permanently abolishes vasoconstriction and sweating and is the best prevention against gangrene. It does not prevent the development of new lesions.

15. **Anticoagulants.** Treatment is begun with heparin and Dicumerol at the same time, and heparin is discontinued when the prothrombin time is sufficiently elevated. These measures are used only during an acute episode, because reflex arterial spasm distal to the point of occlusion is a favorable site for thrombosis.

16. **Local treatment of ulcers, gangrene, infections.** As little as possible is done. The intact skin in the neighborhood of the lesion is extremely vulnerable. Warm boric soaks will encourage drainage, and a 0.05 per cent solution of tyrothricin may be applied.

17. **Remove foci of infection.** This has a favorable effect in reducing arterial spasms.

18. **Surgical treatment of intermittent claudication.** Weakening the muscle affected by the pain reduces its oxygen demand and lessens ischemic pain. An internal popliteal neurectomy divides branches of supply to the calf muscles. An external popliteal neurectomy paralyzes the foot dorsiflexors, which the patient accepts in preference to the pain.

Subcutaneous tenotomy of the Achilles tendon is probably best. It weakens rather than paralyzes and effectively relieves pain in many cases. One must be reasonably certain that the inadequately nourished foot will tolerate surgery.[10]

19. **Amputation.** A gangrenous toe must be allowed to slough spontaneously. Separation may take a long time, but conservatism is safer. After a surgical attempt to remove a toe, the wound may not heal, and gangrene may extend.

When a large portion of the foot appears to be lost, it should be refrigerated while awaiting surgery. Absorption of toxic substances is prevented thereby, and the mortality rate is greatly reduced.

Amputation through the foot rarely heals. The sites of election are 6 inches below the knee or the lower third of the thigh. A histamine flare test may indicate the proper level, but the final determination is made at surgery by noting the amount of bleeding and the condition of the tissues. (See section on Amputations.)

20. **Miscellaneous procedures.** The following measures are described, although their effects are controversial:

A. *Intermittent venous compression.* Theoretically, this causes venous congestion which, when released, may encourage arterial and capillary flow. The mechanical compression can seriously traumatize already damaged vessels.

B. *Intermittent suction and pressure* (pas-

[10] Boyd, A. M., Ratcliffe, A. N., Jepson, R. P., James, G. W. H.: Intermittent claudication, J. Bone & Joint Surg. 31B 325, 1949.

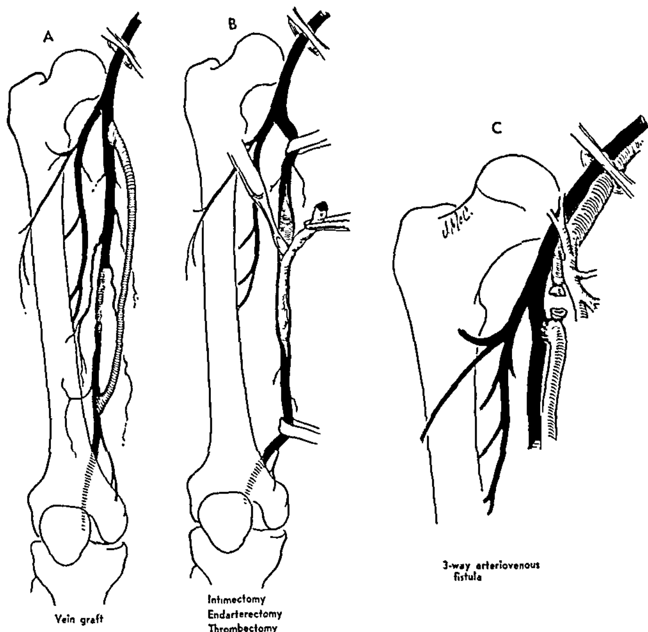


FIG. 255. Surgical procedures for overcoming occlusion of a large artery in an extremity. Such surgery is especially reserved for a recent occlusion in a vessel affected by arteriosclerosis obliterans, the distal small vessels being relatively unaffected. (Redrawn from Pratt, G. H.: Cardiovascular Surgery, Philadelphia, Lea & Febiger)

sive vascular exercise—"Pavex"). The limb is encased in a boot which by alternate suction and compression supposedly opens blood channels and encourages blood flow. It may produce dilatation of blood vessels and capillary atony and is definitely contraindicated in thrombophlebitis and ulcerative lesions.

C. *Tissue extracts.* Occasionally, claudication seems to be abolished by pancreatic extract. Five cc. of deproteinated pancreatic tissue extract is given intramuscularly daily for about 2 weeks, then twice weekly for 4 weeks, and finally once weekly for 4 weeks.

D. *Vitamin E* in high dosage (400 mg. daily) supposedly improves the nutritional appearance of feet.

E. *Intravenous hypertonic sodium chloride solution.* A low incidence of amputations with this treatment has been reported.[11] 300 cc. of a 3 to 5 per cent solution is given intravenously 3 times weekly for 6 months, then twice

[11] Silbert, S.: Thromboangiitis obliterans: results of treatment with repeated injections of hypertonic sodium chloride, J.A.M.A. 94:1730, 1930.

a week for 6 months, and finally once weekly for 6 months. Decreased blood viscosity and increased blood volume is the explanation of its effect. A disadvantage is thrombosis at the site of injection.

DIRECT SURGICAL ATTACK UPON VESSELS[12]

It is possible to remove an obstructing lesion or detour the flow of blood around the point of occlusion This is especially true in cases of arteriosclerosis obliterans where arterial occlusion most frequently takes place in the femoral and the popliteal arteries, whereas the distal smaller vessels are relatively uninvolved. The most desirable situation is a relatively recent occlusion in a young individual with a minimum of arterial disease. Aortography and arteriography localize the point of obstruction A sympathectomy is a necessary preliminary procedure.

1. **Replacement Grafts.** The saphenous vein or the femoral vein may be used as a graft to by-pass the occluded segment. The necessary requisites are:

A Arteriographic evidence of a local block.

B. Remaining vessel is healthy and will hold sutures.

C. Removal of thrombus does not leave an adequate channel.

D. Inadequate collateral circulation.

E. Probability of gangrene without this procedure.

F. Patient is young

PROCEDURES.[12, 13, 14] The segment is bridged without destroying the collaterals. Therefore, the blocked arterial segment must not be removed. The saphenous or the femoral vein is removed, reversed in direction (because of the valves), and its ends are anastomosed to the side of the artery above and below the blocked artery. The artery is opened, a window is excised, and the end of the vein is sewed over with continuous No. 00000 arterial silk. The vein must be kept moistened with heparin-saline solution during the procedure.

2. **Intimectomy, Thrombectomy, Endarterectomy.**[15, 16] In recent years, these procedures are rendered more successful perhaps because of anticoagulants. Heparin can be dripped through a polythene tube inserted above the site of surgery. This type of surgery has an extremely limited field of usefulness. Requisites for intimectomy or thrombectomy are:

A. Traumatic thrombosis in a normal limb.

B. An acute area of thrombosis, especially with undermining of an atheromatous placque.

C. Failure of conservative treatment.

D. Signs of progressive inadequacy of circulation in the extremity and previous loss of the opposite extremity.

E. Thromboangiitis obliterans in young individuals with failure of conservative therapy. (This indication of Gerald Pratt may be questioned in view of the fact that Buerger's disease involves the larger arteries late after the smaller arteries in the leg are extensively involved.)

F. Arteriogram or aortogram shows a localized obstruction.

Contraindications are gangrene, spreading infection, extensive involvement, and thrombosis of the bifurcation of the aorta.

PROCEDURE. The obstructive site is localized by aortography or arteriography. Spinal anesthesia is recommended. The artery is incised over the obstruction. A point of cleavage is found between the inner hard sclerotic cuff containing calcium deposits and the outer adventitia. The internal substance appears like a sequestrum. The external coat is sutured with fine arterial silk. Clotting is prevented by anticoagulants. The inner lining soon endothelializes. Surgical sympathectomy should precede the operation.

When clinically one can determine that an acute occlusion has occurred, the block is localized by aortography. The thrombus, if mov-

[12] Pratt, G H. Cardiovascular Surgery, Philadelphia, Lea & Febiger, 1954.

[13] Leriche, R., and Kunlin, J. Possibilité de greffe veineuse de grande dimensions 15 á 47 cm) dans les thromboses artérielles etendues, Compt rend Acad sc 227.939, 1948.

[14] Kunlin, J., Bitry-Boëly, Volnië, M , and Beaudry, M.· Le traitment de l'ischémie artéritique par la greffe veineuse longue, Revue de Chirurgie, 206-235, juillet-août, 1951.

[15] Dos Santos, J. C . Note sur la désobstruction des anciennes thromboses artérielles, Presse méd. 39 544, 1949.

[16] Bazy, L., Huginer, L , Reboul, H., and Lanbry, P : L'endarteriectomie pour arterite oblıterante des membres inferieurs, J. Int. Chir. 9.45, 1949.

able like an embolus, is removed. If the thrombus is fixed and attempted removal might invite further clots, resection of the arterial segment is advised.

3. **Creation of Arteriovenous Fistulas.** An arteriovenous shunt encourages the development of tremendous collateral circulation in an effort to get blood to the periphery. A 3-way lateral fistula can be created by anastomosing the superficial femoral artery to the distal component of its accompanying vein. Theoretically, and in experimental animals, such a fistula has an adverse effect upon the heart and the kidneys. Therefore, it should be reserved for those cases in which gangrene is imminent and no one point of occlusion can be localized. Approximately 12 to 20 per cent of cases can be salvaged by this therapy.[17] If successful, the fistula probably should be closed later.

ANGIOSPASTIC CONDITIONS

Spasm of an artery or a segment of an artery is a feature of many diseases and syndromes. Active arterial contraction occurs in response to a tactile, thermal, emotional, or chemical stimulus. Two theories explain the mechanism of spasm:

1. **Neurogenic Theory.** The stimulus acts centrally to effect a nerve impulse which excites spasm peripherally.

2. **Chemical Theory.** As a result of the stimulus, adrenalin, sympathin, or some unknown substance called "spastin" is liberated into the blood stream and acts directly upon the arterial muscle.

Arterial spasm may be part of a condition without an organic basis (e.g., Raynaud's disease) or may complicate occlusive arterial disease (atherosclerosis, thromboangiitis obliterans, arterial emoblism). Regardless of the initiating cause, unrelieved spasm can develop into a permanent organic occlusion. When arterial spasm is linked with a definite etiologic factor, sometimes it is called "secondary Raynaud's phenomenon" to distinguish it from the primary or functional type of Raynaud's disease.

[17] Winfield, J. M., and Ruggiero, W. F. Evaluation of arteriovenous shunt and the treatment of its ischemic extremity due to arteriosclerosis, Meeting of the New York Surgical Society, Feb. 1951.

RAYNAUD'S DISEASE[18]

Raynaud's disease is characterized by recurrent attacks of spasm of peripheral arteries, especially the arterioles, without organic basis, predominantly and symmetrically affecting the upper extremities of women. Color changes accompany each attack, and eventually, after months or years, trophic skin changes appear.

Etiology

The actual cause is unknown. Predisposing factors are:

1. **Sex and age.** Women between the ages of 18 and 35 are favored in a ratio of 5:1.

2. **Nervous temperament** is characteristic.

3. **Stimuli** which most frequently excite spasm are cold and emotional stress.

4. **Endocrine relationship** is suggested by the frequent association with menstrual dysfunction and relief with restoration of normal menses.

Symptoms

On exposure to the stimulus the fingers become blanched and waxy, especially at their tips, accompanied by a sensation of numbness —the "dead finger phenomenon." After a variable period, the spasm relaxes, cyanosis may appear, and then the fingers develop a rubor, a bright red hue associated with an intense paresthesia of tingling, "pins and needles," or even pain. The frequency of attacks varies from many times a day to very infrequently. At first the color changes affect only the fingertips. Later, the changes extend more proximally to involve the hands.

If the attacks continue over a number of months, trophic changes develop. The skin becomes thin, smooth and glistening and easily traumatized. The subcutaneous tissue atrophies and hardens so that the finger tapers toward the tip. Extremely painful ulcers and small areas of necrosis may develop over the fingertips. The lesions are often symmetric. Between attacks the hands have a cyanotic hue. Sclerodermatous skin changes may permanently restrict use of the fingers.

[18] Raynaud, A. G. Maurice: DeL'asphyxie locale et de la gangrene symetrique des extremities, p. 177, Paris, Rignoux, 1862.

Pathology[19]

The early pathology is unknown. In advanced stages, intimal thickening and thrombotic occlusion in digital arteries cause the small regions of ulceration and necrosis at the fingertips.

Physiology

Spasm of the arterioles prevents blood from entering the capillaries, observed clinically as pallor. Eventually, the capillaries and the venules become widely dilated, and reflux of blood may occur from the venules to the capillaries where the stagnant blood appears as cyanosis. Either the vasomotor system is at fault, as evidenced by relief by sympathetic ganglionectomy,[20] or an abnormal sensitivity to direct stimuli exists in the digital arteries.[21]

Diagnosis

The main clinical features include predominance in the female, young age, bilateral and symmetric involvement of the upper extremities, and response to exciting stimuli, especially cold. The diagnosis is confirmed by exposing the part to cold and noting the color changes.

A sympathetic block should relieve spasm. The oscillometer and the arteriogram may help to rule out occlusive arterial disease. Postural color changes and absent arterial pulsations are indicative of organic disease being primary and Raynaud's phenomenon as secondary. The cervicobrachial area is examined especially for a cervical rib or scalenous compression.

Treatment

Prophylaxis is mainly the avoidance of the exciting stimulus. General-acting vasodilating drugs as the nitrites, alcohol and papaverine are occasionally helpful in aborting an attack. Local vasodilatation is effected by "Mecholyl" given by iontophoresis. Usually, simple warming of the hands and the body are effective in terminating the attack. In mild cases associated with menopausal symptoms or aggravated during the menstrual period, relief of symptoms may follow administration of estrogens Smoking is prohibited. Repeated sympathetic blocks are of temporary value in an acute phase and will indicate the effect of a sympathectomy. Surgical treatment is indicated only in those cases with progression of symptoms and threatened trophic changes. Surgical resection of the second and the third thoracic ganglia, their rami and accompanying intercostal nerves is advised.

When Raynaud's syndrome complicates an organic arterial disease, the factor of spasm may adversely affect the outcome. Even in the absence of clinically demonstrable spasm, sympathectomy will effect capillary dilatation and should be done wherever possible.

Gangrene generally never affects more than the tip of the finger. Rarely is amputation indicated, and then only a portion of the finger finger is removed.

TRAUMATIC ARTERIAL SPASM

An artery is capable of active sustained contraction in response to trauma acting upon, or in the vicinity of, the vessel. The trauma is either a single, severe blow, acutely affecting a single large artery, or a frequently repeated series of minor traumata, chronically affecting many of the minute vessels. The degree of arterial constriction is greater than that produced by sympathetic stimulation alone. Therefore, the mechanisms by which traumatic arterial spasm is brought about are (1) direct trauma to the adventitia, and (2) reflex sympathetic stimulation.

Chronic Traumatic Arterial Spasm

Frequently repeated minor blows occur in certain occupations such as those requiring the use of a pneumatic tool.[22, 23] The traumata are inflicted mainly in the hands where, due to constriction of the arterioles, Raynaud's phenomenon appears. Attacks of pallor fol-

[19] Spurling, R G, Jelsma, F., and Rogers, J. B. Observations in Raynaud's disease with histopathologic studies, Surg, Gynec. & Obst 54 584, 1932.

[20] Adson, A W, and Brown, G E The Treatment of Raynaud's disease by resection of the upper thoracic and lumbar sympathetic ganglia and trunks, Surg, Gynec & Obst 48:577, 1929

[21] Lewis, T Vascular Disorders of the Limbs, p 111, New York, Macmillan, 1936

[22] Hardgrove, M. A F., and Barker, N. W.: Pneumatic hammer disease, Proc. Staff Meet., Mayo Clin 8 345, 1933.

[23] Drenckhahn, C. H: Vasospastic disease of the hands of miners due to vibration, Illinois M J. 70 354, 1936.

lowed by cyanosis, numbness, paresthesias, pain and perspiration usually are confined to one hand and are precipitated by cold, emotional stress and occasionally by the use of the tool. Attacks are pronounced in the morning or late in the day. When Raynaud's syndrome appears in a male, a source of trauma should be sought.

Treatment. The occupation must be changed, vasodilatation secured by warmth and alcohol, and vasoconstricting influences removed, namely tobacco, cold and excitement. The outlook for improvement is excellent once the cause is eliminated.

Acute Traumatic Arterial Spasm (Arterial Segmental Spasm; Arterial Stupor)

The most common cause is a fracture occurring at certain situations where the artery is bound closely to the bone. An example is at the elbow where the artery may be angulated over and compressed against the upper fragment of a supracondylar fracture of the humerus. On exposure, the artery and frequently its accompanying vein are found to be contracted down to a matchstick size. Spasm of a large artery is associated with widespread spasm of the collaterals distal to the point of trauma. No pulsation is palpable. and the distal end of the limb is pale or cyanotic, cold, anesthetic and painful. After arterial spasm has persisted for some time, muscular weakness and paralysis develop distally.

Acute arterial spasm (and collateral spasm) is rarely sufficient to cause gangrene. However, persisting relative ischemia thus produced causes parenchymatous necrosis and scar tissue replacement of muscles, particularly those in the forearm. (See Volkmann's Ischemic Contracture.)

Treatment. The relief of spasm of a large artery constitutes a surgical emergency. In certain situations, one recognizes the possibility of an embolus or a thrombus occluding the vessel. Although differentiation between these conditions is impossible, it is unnecessary because treatment is almost identical. Obstruction of the main vessel in itself may be harmless, provided that the collaterals are opened.

Delay for a few hours is permissible to observe the effects of nonsurgical measures, reduction of the fracture, lessening the angle of the adjacent joint (especially important at the elbow), sympathetic block and vasodilators (alcohol, papaverine intravenously). Anticoagulants are started immediately inasmuch as thrombosis often follows spasm.

Failure to restore circulation within a few hours is sufficient indication for exploration. Overlying fasciae are resected, the fracture is reduced under direct vision, and the vessels are freed. A warm solution of normal saline containing a local anesthetic is poured into the wound to bathe the vessels. If vasodilatation cannot be secured in this manner, it is advisable to resect the affected segments of both vessels. Ordinarily, this removes the reflex constriction of the collaterals. Nevertheless, a sympathectomy must follow this procedure.

Where resection of an arterial segment would threaten viability of the part (as in organic occlusive disease), alternatives are (1) resection with end-to-end anastomosis, (2) resection and artery or vein graft, or (3) bridging with a graft without resection. (See Acute Arterial Injuries.)

SUDECK'S ATROPHY (Reflex Sympathetic Dystrophy; Post-traumatic Painful Osteoporosis)

In 1900 Sudeck[24] first described an acute atrophy of bone with characteristic spotty decalcification, developing after trauma, and associated with pain, edema, tenderness, cyanosis, coldness, sweating, and stiffness of the part. He attempted to define a specific disease entity. However, these changes are a result of vasospasm of terminal portions of the arterioles which develops in response to various stimuli and is found in a variety of conditions, usually traumatic in origin. Therefore, Sudeck's type of spotty osteoporosis is one manifestation of a physiologic response of the sympathetics to an irritative focus.

Physiologic-Pathologic Mechanism

The irritative stimulus, often traumatic in origin (fracture, incomplete nerve injury,

[24] Sudeck, P.: Über die akute (reflektorische) Knochinatrophie Nach Entzündungen und Verletzungen An den Extremitaten und ihre klinischen Erscheinungen, Fortschr. Geb. Röntgenstrahlen 5:277, 1901-1902.

Clinical Picture

The onset of pain varies from immediately after injury to several weeks later. The injury is often trivial in nature, such as a sprain, although any type of trauma is causative. The more severe injuries most frequently are those which cause a partial lesion to a peripheral nerve. The pain is constant, intense and burning; rarely, is it knifelike, crushing, paresthetic, etc. It is distributed diffusely over the distal portion of a limb not related to a nerve distribution, frequently over the palm of the hand and the plantar aspect of the foot. Various stimuli which intensify the pain include touching or tapping the part, noises, dependent position, emotional disturbance, etc. Usually the discomfort is worsened by dryness and heat and eased by moisture and coolness; not uncommonly the opposite effect is obtained. The skin over the affected area is exquisitely hyperalgesic. Within the first few weeks the appearance of the part is normal, but eventually manifestations of autonomic disturbance appear. The skin becomes reddened, blotchy, dry, edematous and warm, a picture of vasodilatation. The skin temperature is elevated. Later, sometimes without the antecedent vasodilatation, the skin becomes cold, pale, cyanotic, perspiring, thin, glossy, and nails are brittle and ridged. Oscillometric readings are usually equal to those in the uninvolved extremity. This is surprising when vasodilatation and elevated skin temperature is present. This suggests that interruption of sensory nerves can cause only surface vasodilatation and not constriction or dilatation of the larger vessels.

Diagnostic Sympathetic Block

Interruption of sympathetic impulses by infiltration of local anesthetic about the sympathetic chain will reduce or completely eliminate the pain. For the upper extremity, the needle is inserted between the first and the second ribs. For the lower extremity, the anesthetic is injected adjacent to the first and the second lumbar vertebrae. Failure to obtain relief may be due to improper technic, and the procedure should be repeated.

Course

With passage of time, the pain may spread upward or to the opposite limb The patient becomes emotionally unstable, hyperirritable and, if the pain persists, may even develop suicidal tendencies. It is said that if the pain is allowed to spread centrally, it may become permanently implanted in the central nervous system and completely unresponsive to treatment.



Fig. 256. Reflex sympathetic dystrophy. The spotty osteoporosis distal to the fracture site is typical.

Etiology and Pathologic Physiology

The cause is usually trauma, which is often trivial in nature. In severe injuries the pathologic feature is a partial lesion of a peripheral nerve, most commonly of the median or the sciatic nerve or the brachial plexus. When paralysis is complete and associated with causalgia, one finds at surgery a neuroma with the nerve in continuity; electric stimulation of this nerve fails to evoke a motor response.[25] Never does division and separation of nerve ends occur.

Sympathetic dysfunction is evidenced by vasodilatation and vasoconstriction. Various theories have been advanced to explain the mechanism. A plausible theory states that efferent sympathetic impulses are shunted into sensory fibers of a mixed nerve. This explains

etc.), reflexly provokes continuous vasospasm of the terminal arterial channels. Larger vessels remain unaffected so that oscillometric readings remain unchanged. Because vasospasm and its resultant changes usually develop around and distal to the site of injury, irritation of sympathetic fibers in the peripheral nerves and the perivascular coats seems highly probable. Capillaries and venules become distended with sluggishly flowing blood, lacking the propulsive pressure transmitted from the arterioles. This explains cyanosis and coldness of the part. Slowly moving blood develops a low pH which promotes dissolution of mineral salts of bone. The spotty arrangement of decalcification may be explained by a similar distribution of capillary beds within the bone. *Arteriolar spasm effects a relative* ischemia of all tissues. As a consequence, increased permeability of capillaries and venules permits diffusion of plasma and its fibrin content into the tissue spaces, manifest clinically as edema. Fibrin deposits are followed by fibrous tissue replacement. Finally, as vascular channels are reopened, bone minerals are redeposited. Fibrosis throughout the soft tissues retards vascularization so that color and temperature are slowly restored. In addition, mobility of joints is restricted and never fully recovered. Hyperhidrosis is a result of overstimulation of the sympathetics. All the described changes may be appropriately termed "a reflex sympathetic dystrophy."

Clinical Picture

Trauma varying from a trivial to a severe injury precedes the appearance of symptoms. The injury often occurs about a joint, especially the wrist and the ankle. The following changes develop in the hand or the foot: swelling, edema, tenderness, cold, moist, slightly cyanotic glossy skin, and limitation of motion of the digits. Pain on movement of the part encourages immobilization and further restriction of motion. When pain becomes intense, persistent, often burning in nature, and aggravated by certain stimuli (heat, emotion, touching the part), this symptom becomes the eminent part of the syndrome which is then designated as *causalgia* (q.v.). Symptoms and findings in the average case last a period varying from weeks to months, then gradually subside, leaving a hand or a foot which is stiff, cold, slightly cyanotic. In the hand, fibrotic contracture of the metacarpophalangeal joints in the extended position constitutes a severe disability.

Roentgenologic Findings

Spotty decalcification develops throughout bones distal to the site of injury. For example, in a fracture of the lower third of the tibia, osteoporosis develops in the distal fragment and all the bones of the foot. A diffuse deossification of disuse may be superadded.

Treatment

During the actively vasospastic phase, blood flow is encouraged by elevation and active exercise of the extremity. In the hand, attention is directed to preserving the full range of motion in the metacarpophalangeal and the interphalangeal joints. When pain is severe and persistent, and marked swelling threatens function, sympathectomy will effect rapid recovery. This surgical procedure must be performed early before fibrotic contractures have supervened. Persistent stiffening of the metacarpophalangeal joints in extension will require *multiple capsulotomies.*

The pain of causalgia is due frequently to a partial nerve injury. Relief may require not only sympathectomy but also resection and repair of the traumatized nerve. (See Causalgia.)

CAUSALGIA[25-28]

Causalgia is a condition characterized by post-traumatic pain which is persistent, diffuse and burning, occurring in paroxysms, and provoked by various stimuli. Its relief by interruption of sympathetic impulses classifies it as a sympathetic dystrophy, and as such it *is related* to other conditions of sympathetic origin, for example, Sudeck's atrophy, trophic edema and reflex arterial spasm.

[25] Shumacker, H. B., Spiegel, I. J., and Upjohn, R. H.: Causalgia, Surg., Gynec. & Obst. 86:76, 452, 1948.

[26] Mitchell, S. W., Moorehouse, G. R., and Keen, W. W.: Gunshot and Other Injuries of Nerves, Philadelphia, Lippincott, 1864.

[27] Mayfield, F. H.: Causalgia, Am. J. Surg. 74:522, 1947.

[28] Miller, D. S., and deTakats, G.: Posttraumatic dystrophy of the extremities, Surg., Gynec. & Obst. 75:558, 1942.

Pathologic Changes

On exposure to cold, vasoconstriction takes place, principally in the arterioles, and may persist for a day or more. In mild frostbite a low-grade inflammatory reaction takes place in the vessels and the surrounding tissues. On prolonged exposure to cold and persistent vasoconstriction, inflammation develops in the intima of small arteries and arterioles. The endothelium of capillaries is damaged, permitting abnormal permeability. In the process of thawing, on exposure to higher temperatures, a reactive hyperemia occurs. Thrombi form in the terminal arterioles, and blood and plasma extravasate through the damaged capillary walls, causing edema and blood-filled blisters. The fluid accumulation is rather superficial, splitting the epidermis and the dermis. Superficial necrosis develops as the increased oxygen requirements by the normal tissue are not fulfilled by the inadequate blood supply.

Clinical Picture

Frostbite can be classified according to the severity of tissue changes.

Mild Form (First-Degree). With vasoconstriction a dull pallor develops in the skin, accompanied by numbness and a prickling sensation. On warming the part, mild erythema or normal color returns without damage to the tissues. The affected area may remain unusually hypersensitive to subsequent exposure for a considerable time.

Moderate Form (Second-Degree). Exposure to cold of a more extreme degree or for a longer period causes actual solidifying by freezing of the part, which appears white, rigid and insensitive. On thawing, a severe reactive hyperemia develops, starting in the vicinity of, and gradually spreading over, the ischemic, pale skin. The entire involved part is red, tender, edematous, and blisters soon appear. Burning pain and paresthesias are often intense. Superficial sloughs of skin occur, and the part recovers.

Severe Gangrenous Form (Third-Degree). The appearance is that of the second-degree type, but it progresses to gangrene, usually of the superficial tissues. Gangrene is more likely to develop when exposure to severe cold is prolonged, trauma is inflicted (e.g., an enforced march), or the part is kept immobile. Thrombosis develops and, upon warming of the part, gangrene appears.

Treatment

Prophylactically, the main factors to be emphasized are:

1. Clothes loose, light, warm and windproof
2. Feet should be kept dry and oiled.
3. Daily hygiene of feet
4. Moderate activity is desirable; prolonged marches in cold weather are avoided.

Active treatment aims at relief of ischemia as a vascular emergency. Mild frostbite requires only warming by any means at hand. Avoidance of trauma, e.g., vigorous rubbing of the affected part, and maintenance of asepsis are mandatory to prevent more severe involvement.

Severe degrees of frostbite require even stricter adherence to the principles of atraumatism and asepsis. The affected part is treated with utmost gentleness. All compressing and constricting clothing is removed, and the involved region is exposed to the air but protected with a cradle. Smoking is strictly prohibited. Heparin and Dicumarol are started immediately to combat thrombosis and possibly reduce the incidence or the degree of gangrene.[29] Antibiotics are administered.

A controversy exists as to the benefits of maintaining a warm environment to warm the part rapidly, or a cold environment with slowly rising temperature so as to warm the part gradually.

Slow-Warming Method. This is based upon the fact that the venous and the lymphatic vessels suffer the greatest damage and are incapable of draining the blood brought in by the newly opened arteries. Blebs, edema, ecchymosis and gangrene are evidence of excessive fluid supply. If the arteries are only partially dilated by maintaining the part in a cool tent (55° to 60° F.), drainage of blood from the area is adequate. Ice bags, which cool the tent, are gradually decreased over a period of several weeks. The patients are gen-

[29] Lange, K., and Boyd, L. F.: The functional pathology of experimental frost-bite and the prevention of subsequent gangrene, Surg., Gynec. & Obst. 80 346, 1945.

the fact that anesthetic blocking of posterior root inflow will abolish causalgia without effecting sympathetic outflow. The spread of pain probably takes place in the internuncial pool.

Treatment

Very rarely does the condition subside spontaneously. When it does so, the diagnosis is in doubt.

Conservative. Warm applications often help the vasoconstrictor type; cool wet applications may relieve discomfort of the vasodilated type. Rest and elevation reduce the hyperemia and the edema. Preganglionic blocking agents are effective occasionally:

Tetraethylammonium chloride (Etamon Chloride, Parke-Davis) is given intramuscularly every 6 hours in a dose not exceeding 20 mg per Kg. of body weight; the block should not be continued for more than 36 hours. Contraindications are severe hypertension, impaired renal function, a high diastolic pressure and a recent coronary occlusion.

Benzazoline hydrochloride (Priscoline Hydrochloride, Ciba) is given orally in a dosage up to 50 mg. 6 times daily. Because it also has a local effect in causing vasodilatation, it may accentuate symptoms in the vasodilated type of causalgia.

Nylidrin Hydrochloride (Arlidine, Arlington-Funk). This agent has a minimum of side-effects. Its dosage is 6 m.g. t.i d.

Procaine injections of the sympathetic trunk are effective in many cases. It is done daily, and the relief obtained varies from an hour to permanent cure even on one injection. For the upper extremity, 0.5 per cent procaine with Adrenalin is injected into the paravertebral space between the first and the second dorsal vertebrae. An effective block is indicated by the Horner's syndrome, cessation of sweating and vasodilatation. For the lower extremity, the injection is made between the first and the second lumbar vertebrae

Surgical treatment consists of sympathectomy. When done at an early age, the results are almost invariably good, the pain being greatly reduced or entirely eliminated. Before surgery, it is necessary to perform a procaine block to note the effect of interrupting sympathetic impulses. Delay in performing surgery is unwise, as permanent trophic changes and deformity may develop.

Upper extremity causalgia is treated by sectioning the thoracic trunk below the third ganglion and removing the gray and the white rami to the second and the third ganglia.

Lower extremity causalgia requires removal of several upper lumbar ganglia, from the first to the fourth. Results are better when the first lumbar ganglion is included

When a partial nerve lesion can be identified, neurolysis, resection of the neuroma and neurorrhaphy are necessary in addition to sympathectomy in order to eliminate residual partial pains and paresthesia. These procedures in themselves are of no value in eliminating the severe causalgic pain. Very late cases of causalgia rarely require cordotomy or lobotomy, but the results leave much to be desired.

FROSTBITE

Frostbite is defined as ischemic destruction of superficial soft tissues as a result of exposure to cold. Vasoconstriction plays a prominent part initially, but arteriole thrombosis and increased capillary permeability cause later damage.

Etiology

Cold produces vasoconstriction locally and reflexly. The resultant ischemia, if severe and prolonged, leads to thrombosis of the arterioles with gangrene. Death of tissue is also caused by direct freezing to a solid state of cellular fluids. Although the true freezing point of the skin is between −2° C. (28.4° F.) and 0° C. (32° F.), the skin has the ability, because of the phenomenon of supercooling, to go below its freezing point without solidifying. Therefore, freezing of the skin does not occur until −10° C. (14° F.), or lower. The application of oils to the skin increases the capacity for supercooling.

Factors contributing to freezing so that pathologic changes develop at a higher temperature are: (1) *moisture,* (2) *wind,* (3) *inactivity* (immobility encourages venous stasis), (4) occlusive arterial disease, (5) tobacco excess and (6) anoxemia (high altitude flying, anemia).

and separates as healthy granulations form beneath the crust. The final result is an atrophic, contracted foot which appears dusky, cold and hyperhydrotic as manifestations of sympathetic overactivity in response to cool temperatures.

TREATMENT

Prevention demands avoiding exposure for prolonged periods to cold and wet, encouraging activity, and frequent changes of shoes and socks.

General treatment includes antibiotics, prohibition of smoking, and administration of anticoagulants.

Local treatment aims at re-establishig vasomotor tone and keeping the tissues at a low metabolic level in the hyperemic stage. The legs must be elevated at intervals to reduce edema, which may interfere with circulation directly or at a later time by resulting scarring. Probably it is best to keep the feet cool by a blowing fan or refrigeration with ice bags until vasodilatation can be overcome. Sympathectomy is not indicated at this stage, because the capillaries and the venules are incapable of handling an increased inflow of blood.

Surgical restraint is adhered to strictly during the process of separation of the superficial gangrenous crust. Later, the granulating surface may be covered with skin grafts

The sequelae, namely, such symptoms as paresthesias, anesthesias, cold sensitivities, a feeling of the foot being cold, and hyperhydrosis, are symptoms of sympathetic overstimulation. The circulation may be improved by a warm climate, avoidance of smoking, typhoid vaccine, etc, but surgical sympathectomy is best.

When frostbite, trench foot, or immersion foot develops at temperatures far above those at which changes might be expected to occur, arterial occlusive disease should be suspected. Gangrene is generally more likely, extensive, and often requires amputation.

PERNIO

(Chilblains; Dermatitis Hemialis)

Pernio is a vasospastic disease, affecting the smaller vessels of the skin and resulting in necrosis and ulceration.[31] Although the chief manifestations affecting the skin of the leg, and less commonly the dorsum of the foot and the hand, cause the case to come under the care of the dermatologist, eventually the cause is traced to exposure to cold and wet. Sympathectomy results in a complete cure.

ACUTE PERNIO

Often seen in children, exposed areas of the legs develop multiple spots of dermatitis which are reddened and slightly raised. Intense burning and itching characteristically are aggravated by exposure to warmth. The lesions are bilateral and symmetric in distribution and clear up in about 10 days leaving spots of pigmentation.

CHRONIC PERNIO

On repeated exposure to cold, recurring crops of cutaneous lesions appear, but they disappear in the warmer months. Red, elevated, painful, small lesions appear on the lower portions of both legs anteriorly and posteriorly, accompanied by an itching and burning sensation. On the surface of the erythematous lesions, blisters form and break down, leaving hemorrhagic ulcers, each with a violaceous base. Pain subsides with appearance of ulceration. The ulcerative lesions heal in 3 to 5 weeks, leaving permanent scarring and pigmentation. No clinical evidence of arterial occlusive disease can be found, but biopsy of the lesions will reveal angiitis with low-grade inflammation in the subcutaneous tissue. Absence of tubercles rules out Koch's infection.

TREATMENT

Acute and chronic pernio may be prevented by a continual warm environment. An acute lesion requires protection from trauma and infection because the local area is ischemic. Adequate clothing covering the legs may reduce the possibility of recurrence.

Chronic pernio is often typified by progressively increasing susceptibility to cold. Eventually, the lesions which at first develop only during the cooler months may appear also after several years, during cooler days of summer months. Treatment is directed toward increasing peripheral arterial circulation. Administration of typhoid vaccine intravenously, or acetyl-β-Methylcholine chloride (mecholyl chloride) by iontophoresis, may be tried. A

[31] McGovern, T., and Wright, I. S · Pernio; a vascular disease, Am Heart J 22 583, 1941.

erally more comfortable in cool surroundings, and the amputation rate by this method is extremely low.

RAPID-WARMING METHOD. Theoretically, warming a part to the point where its increased metabolic demands cannot be satisfied by inadequate oxygen supply results in death of tissue. This explains why, on exposure to cold, gangrene is more likely under anoxemic conditions However, experimental work on animals has demonstrated that rapid warming of the part, but not heating, resulted in no more tissue loss than slow warming.[30] The procedure merely requires immersing in mildly warm water.

After the first week, the degree of tissue destruction will become apparent. The black, dry gangrenous tissue becomes sharply delimited and should be allowed to separate and slough spontaneously. A "hands-off" surgical policy is best The gangrene is superficial, involving mainly the skin and the superficial tissues and, when shed, leaves a bed of healthy granulations which can be covered with skin grafts. Surgical trauma imposed prematurely will only intensify the extent of destruction. While awaiting spontaneous separation, sterile dressings, antibiotics and anticoagulants are continued Accumulation of infective material will usually drain in a warm saline soak.

Sympatheticolytic drugs and surgical sympathectomy may be beneficial in cases with antecedent organic arterial occlusive disease.

TRENCH FOOT AND IMMERSION FOOT

Trench foot and immersion foot are the military and naval counterparts of frostbite. The vascular changes, because of *moisture*, take place at temperatures above those required to cause freezing or frostbite. Moisture acts to carry away the heat more rapidly from the affected part. *Edema* is more pronounced than in frostbite because of prolonged dependency. Under conditions of starvation and hypoproteinemia, edema is greater in degree. Edema implies deposition of fibrin throughout the soft tissues, leading eventually to fibrosis and stiffness of the part. In addition to moisture, *immobility* such as occurs in trench warfare encourages pathologic responses to less than freezing temperatures.

PATHOLOGY

Damage to capillary and venous channels (which may be ischemic in origin as a result of prolonged arteriolar spasm) results in pathologic permeability of the vessel walls. Red blood cell diapedesis and plasma diffusion into the surrounding tissues occur. Thrombosis develops, particularly in venous channels, the clot appearing to be a concentration of red corpuscles rather than actual coagulation with fibrin. Multiple areas of ischemic necrosis develop throughout the soft tissues and are replaced by scarring, particularly in muscle and nerve structures and vessel walls. When necrosis is extensive, it is usually confined to the skin and the superficial tissues.

CLINICAL PICTURE

Several stages may be identified:

1. **Vasospastic Phase.** The first symptoms are numbness, coldness and a sensation of "feet made of wood." Pain is usually minimal during the anesthetic period. Both feet are pale or cyanotic, often mottled, and cold. After the boots are removed, swelling occurs, and intense pain and paresthesia develop.

2. **Stage of Hyperemia.** As the extremities are exposed to warmth, the feet become red and hot, edema increases, and blebs containing a hemorrhagic fluid appear. An intense burning pain is often associated. Occasionally, a shooting, stabbing pain characteristic of ischemic neuritis is experienced. This stage lasts about 2 weeks and may heal completely. Frequently, such feet remain somewhat cyanotic and cold and are hypersensitive to ordinary cold weather, the Raynaud's phenomenon being manifest. On the other hand, this stage may progress further to that of frank gangrene.

3. **Stage of Gangrene.** This varies from small blisters and ulcers to gangrene. Although the skin alone appears necrotic, diffuse ischemic degenerative changes followed by fibrous replacement are taking place throughout the deeper tissues. Muscles become fibrotic, and contractures of the joints of the foot develop. The gangrene is usually superficial

[30] Shumacker, H B, and Kunkler, A W Studies in experimental frostbite IX Rapid thawing and

and separates as healthy granulations form beneath the crust. The final result is an atrophic, contracted foot which appears dusky, cold and hyperhydrotic as manifestations of sympathetic overactivity in response to cool temperatures.

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Treatment

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[31] McGovern, T., and Wright, I. S.: Pernio; a vascular disease, Am Heart J. 22 583, 1941.

surgical sympathectomy will heal the lesions promptly and prevent further recurrences.

CONDITIONS CHARACTERIZED BY VASODILATATION

The following described conditions, erythromelalgia and acrocyanosis, display capillary vasodilatation as part of the pathologic physiology. However, this does not imply that dilatation of the minute vessels is the cause of symptoms. In erythromelalgia the reddening and the warmth indicate that the arterial blood supply is normal and, although the capillaries are distended, the rate of blood flow is adequate. Therefore, sympathectomy is of no value. In acrocyanosis, coldness and cyanosis indicate sluggish blood flow Arteriolar spasm satisfactorily explains this condition as evidenced by *diminution of cyanosis during sleep* and after sympathectomy.

These diseases, although not common, must be differentiated from the more common vascular diseases Erythromelalgia must be distinguished from the painful dependent rubor of occlusive arterial disease and from the burning distress of peripheral neuritis. Acrocyanosis must be differentiated from Raynaud's disease and from pulmonary and cardiac causes of cyanosis.

ERYTHROMELALGIA (Erythermelalgia)

Erythromelalgia is a condition characterized clinically by paroxysms of reddening and warming of the skin in an area of an extremity associated with burning distress. Attacks are invariably provoked by heating the extremity. Two types may be recognized.

Primary Type. Idiopathic, no evidence of vascular or nervous disease Usually, both *lower extremities are involved.*

Secondary Type. Usually, part of hypertension or polycythemia vera. Also occurs in gout, organic nervous disease and heavy metal poisoning. As a rule, only one extremity is involved.

Pathologic Physiology

Vasodilatation is the cause of warming of the skin. The skin must be warmed above a certain minimum, the "critical point," usually between 32° and 36° C., in order to induce a burning distress. Symptoms may be provoked more easily by increasing the hydrostatic pressure such as by compressing the veins proximally or by lowering the extremity. Con*versely, elevating the extremity or applying* direct pressure over the painful area will lessen the distress. Incomprehensible is the difficulty of producing pain by immersing the extremity in warm water.

Clinical Picture

Adults are affected most commonly. The hands or the feet are involved and often only a small portion of the foot. The patient may state that painful burning arises while walking or at *night when the foot is placed beneath* the covers. An attack lasts from minutes to hours. Symptoms are more intense in the warm summer months. Relief is obtained by exposure to cold.

Examination during the paroxysm reveals the painful part to be reddened or cyanotic and warm. Some swelling may be noted Trophic changes are absent except in the secondary type. Because of its frequent association with polycythemia vera, an enlarged *spleen should be sought.*

Laboratory Findings

During the attack a sample of venous blood removed from a vein proximal to the reddened area will reveal an increased oxygen saturation. The blood count is checked for polycythemia vera.

Differential Diagnosis

Similar symptoms are associated with ar*teriosclerosis* and peripheral neuritis. However, the skin is not warm, and symptoms are not induced by exposure to heat.

Treatment

No specific treatment is available at present. Ordinary acetylsalicylic acid often gives relief for several days. Marked relief may be obtained from epinephrine Prophylactically, warm environments must be avoided, and light clothing should be worn. During an attack, the extremity is elevated and surrounded by cold packs. Surgically, one may try peripheral nerve section. It may be possible to desensitize the extremity gradually to warmth. The part is exposed to controlled heat which at first is

well below the critical point. Each day the temperature is raised 1° until the critical point is greatly exceeded.

Acrocyanosis

Acrocyanosis is a condition characterized by painless, persistent cyanosis and swelling of the hands and the feet. It occurs most commonly in females. The cause is unknown. The most plausible theory is that of arteriolar spasm causing anoxemic dilatation of the capillaries and the venules. The fact that during sleep the hands of the acrocyanotic patient become warm and red suggests a hyperreactive sympathetic nervous system.

Clinical Picture

The patient is often female. The coldness and the bluish discoloration of the hands and to a lesser degree of the feet have been present for many years. The discoloration is more marked during the winter when the affected parts may be swollen, painful and tender. During the summer the color is less cyanotic. No episodes of blanching occur, and trophic changes, ulceration and gangrene are absent, thereby distinguishing this condition from Raynaud's disease and occlusive arterial disease.

Treatment

No treatment is necessary for this relatively asymptomatic condition. In severe cases sympathectomy is indicated.

ACUTE ARTERIAL INJURIES[32]

A laceration of a major artery may occur during surgery and from other trauma. When the vessel involved is an end artery, such as the popliteal, ligation will be followed by gangrene. Interruption of any large artery at least will cause ischemic changes in soft tissues, such as the development of Volkmann's contracture. The fate of the part supplied depends upon (1) collateral circulation and (2) the site of ligation. For example, ligation of the superficial femoral or brachial arteries will cause gangrene more frequently than ligation of other arteries In these two arteries, beyond their main collateral branch, are segments from which no collateral branches arise. If the artery is ligated at some distance distal to the main collateral branch, a blind arterial pouch remains into which the arterial blood is directed and in which arterial pressure is needlessly dissipated. As a result, the arterial pressure within the main collateral branch is reduced and inadequate, and gangrene frequently develops. *Ligation is done immediately distal to the large collateral branch,* thereby avoiding the blind pouch, and the incidence of gangrene is lessened. In any event arterial repair is preferred to ligation.[33]

When a major normal artery is injured, the damaged artery and the collaterals undergo spasm. This physiologic spasm squeezes out the blood and may be nature's method of reducing hemorrhage.

Etiology of Arterial Injuries

Consideration is given here to wounds, fractures and crushing injuries.

Types of Arterial Injuries

1. Complete Division. Exsanguinating hemorrhage is not always a sequel, because the vessel ends retract, the artery undergoes spasm, and thrombosis seals the lumen.

2. Partial Division. Hemorrhage is severe, and secondary hemorrhage is likely, because the arterial ends cannot retract. A large blood clot may accumulate over the opening, forming a *pulsating hematoma.* The arterial pressure develops a cavitation within the hematoma, while the outer wall of the mass becomes organized, resulting in a *false aneurysm.*

Simultaneous partial division of the accompanying vein allows arterial blood to track from artery to vein. A definite connecting channel forms, and an *arteriovenous fistula* results.

3. Contusion. Spasm not only of the damaged vessel but also of the collaterals frequently follows injury. This type of spasm is intense and only partially relieved by sympathetic block. Thrombosis is not necessarily a sequel, because this requires damage to the intima, plus slowed blood flow.

4. Compression. A bone fragment is the usual cause. The main danger is that of thrombosis. Vascular compression occurs, especially

[32] Freeman, N. D.: Acute arterial injuries, J.A.M.A. 139:1125, 1949.

[33] Holman, E.: Further observations on surgery of the large arteries, Surg., Gynec. & Obst 78 275, 1944.

surgical sympathectomy will heal the lesions promptly and prevent further recurrences.

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Laboratory Findings

During the attack a sample of venous blood removed from a vein proximal to the reddened area *will reveal an increased oxygen saturation.* The blood count is checked for polycythemia vera.

Differential Diagnosis

Similar symptoms are associated with arteriosclerosis and peripheral neuritis. However, the skin is not warm, and symptoms are not induced by exposure to heat.

Treatment

No specific treatment is available at present. Ordinary acetylsalicylic acid often gives relief for several days. Marked relief may be obtained from epinephrine. Prophylactically, warm environments must be avoided, and light clothing should be worn. During an attack, the extremity is elevated and surrounded by cold packs. Surgically, one may try peripheral nerve section. It may be possible to desensitize the extremity *gradually to warmth.* The part is exposed to controlled heat which at first is

wall, or when a rise in blood pressure overcomes resistance of the wall. Thus an emotional upset or a lifting strain by increasing intra-arterial tension may cause an aneurysm to appear suddenly.

Constriction of an artery often causes an aneurysm to develop just beyond the point of narrowing. An example is compression of the subclavian artery by a cervical rib. Theories of pathogenesis include: (1) interruption of sympathetic constrictor impulses to the local segment, (2) abnormal blood currents just beyond the stenosed channel.[36]

ETIOLOGY

Aneurysms are congenital or acquired.

Congenital Arterial Aneurysm. This is rare. It is a result of an underdeveloped or absent muscle layer, the tunica media. It is found most frequently intracranially at the base of the brain where rupture causes fatal hemorrhage.

Acquired Arterial Aneurysm. This results from trauma or disease weakening the arterial wall. In the presence of a diseased vessel, trauma often precipitates the formation of an aneurysm.

1. TRAUMATIC ANEURYSM. Laceration of the vessel is caused from without by a *penetrating wound* or from within by a *fracture*. Only a portion of the circumference of the vessel is involved. When the laceration penetrates only the outer layers, arterial pressure dilates the inner layer. When the laceration extends through all the coats but the artery is held in continuity by part of the wall, there develops a large hematoma which temporarily obstructs the arterial flow. The pulsations of arterial pressure are transmitted by the hematoma and penetrate and enlarge the interior of the soft clot until the blood flows into the distal arterial component. Eventually, the distended clot becomes endothelialized on its inner surface, and the wall is reinforced by fibrous tissue and surrounding structures. The cavity becomes part of the peripheral artery circulation. Laminations of blood clot, as they deposit on the wall, become organized and thicken the wall to a leatherlike consistency.

[36] Holman, E: On circumscribed dilatation of artery immediately distal to partially occluding hand. poststenotic dilatation, Surgery 36.3, 1954.

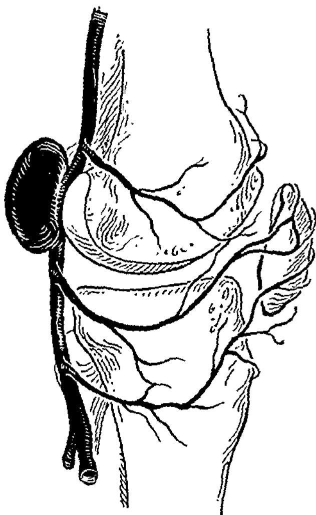


FIG. 257. Popliteal aneurysm.

2. DISEASE CAUSING ANEURYSMS. In peacetime, 80 per cent of aneurysms are the result of arterial disease. Under conditions of war, trauma is the main cause.

A. *Arteriosclerosis* is the commonest cause. A calcium deposit is the usual site of rupture Slight trauma to a sclerotic vessel is often the precipitating cause. The most common sites of saccular arteriosclerotic aneurysms are the popliteal arteries and the abdominal aorta, less commonly the femoral artery just beyond the groin.

The usual site of an arteriosclerotic aneurysm is an artery not surrounded by skeletal muscles and subject to frequent bending. They usually develop after the age of 60, and men are favored in the ratio of 10 to 1. Complete rupture is rare, especially in the extremities. Complete thrombosis often occurs in popliteal aneurysms.

B. *Embolus.* The blood clot or atheromatous

at the elbow or the knee, where the vessels are held fixed close to the bone.

Clinical Picture

Hemorrhage may or may not be in evidence. The distinguishing features are:

1. **Extreme pallor** due to spasm of the artery and its collaterals
2. **Loss of pulsation**
3. **Oscillometry** may confirm loss of pulsation
4. **Glovelike anesthesia and paresis or paralysis** are due to ischemia of nerves and muscles.
5. **Severe Pain**
6. **Auscultation.** A systolic bruit indicates a false aneurysm; a continuous to-and-fro bruit indicates an arteriovenous aneurysm. The distinction between the two is important. In the false aneurysm, arterial pressure is high, and the mass tends to expand; also, it compresses adjacent structures. Rarely, it may rupture. On the other hand, the intrasaccular pressure in an arteriovenous aneurysm is low. The lesion stimulates the development of collaterals; therefore, operation may be postponed advantageously.

Treatment

In most cases diagnosis and treatment are urgent. Delay may result in thrombosis, occlusion of collaterals, and irreversible nerve and muscle changes. Management is outlined as follows:

1. **Immediate Treatment**

A. *Arrest hemorrhage*

a. Avoid tourniquet if possible, because it cuts off collaterals.

b. Secure direct pressure on bleeding point by plain gauze, forceps, or hemostatic gauze (oxycel, gelatin sponge, fibrin foam).

B. *Restore blood volume, hemoglobin, protein.*

2. **Early operative treatment** aims at restoring blood flow through the original channel.

1. *Débridement*
2. *Avoid ligation proximally,* because it needlessly sacrifices collaterals.
3. *Arterial clamps* are placed proximally and distally.
4. *Excise damaged portion* of vessel.
5. *Anticoagulants.* The vessel is irrigated with a solution containing 10 mg. of heparin per 100 cc. of normal saline.
6. *Close defect* with everting mattress suture.
7. *Vein graft replacement,* if destruction is extensive.

3. **General treatment of ischemic extremity** aims at maximum arterial inflow and minimal metabolic demands.

1. *Keep limb horizontal or dependent,* never elevated.
2. *Cool environment* is desirable.
3. *Avoid refrigeration* unless amputation is inevitable.
4. *Carry out sympathetic block or sympathectomy.*
5. *Administer vasodilating agents*—whisky, papaverine, etc.
6. *Employ fasciotomy* to avoid ischemic necrosis of muscles. This procedure is questionable, because insufficient circulation may compromise healing of the operative wound.

ARTERIAL ANEURYSM[34, 35]

Definitions

An arterial aneurysm is an abnormal outpouching from an artery, causing a pulsating tumor. This dilation may occur as a gradual enlargement over an extensive area—the *fusiform* type of aneurysm; or it may be localized to a small segment as an abrupt enlargement —the *saccular* type of aneurysm. If the wall of the sac is made up of the vessel structure itself, it is designated a *true aneurysm.* If the vessel wall is destroyed, and the wall of the sac is composed of a blood clot, fibrous tissue, or surrounding structures, it is termed a *false aneurysm.*

Pathogenesis

The factors necessary for the production of an aneurysm are a *weakened blood vessel wall* (especially the muscular coat) and *increased intraluminal pressure.* A vessel wall which has been weakened by antecedent injury or disease may not develop an aneurysm until *trauma,* even of a trivial type, further weakens the

[34] Pratt, G. H.: Cardiovascular Surgery, Philadelphia, Lea & Febiger, 1954

[35] Allen, E. V., Barker, N. W., and Hines, E. A., Jr.: Peripheral Vascular Diseases, Philadelphia, Saunders, 1946

to calcify, especially in arteriosclerotic aneurysms.

Pressure Effects. The accompanying vein may become occluded and thrombosed. Nerves are destroyed. Bone is eroded. Adhesions interfere with adjacent muscle movement.

Effects on Circulation. Blood supply distal to this area is diminished. The effect is accentuated by a reflex spasm of collateral vessels, or an embolus thrown off from the aneurysmal clot.

Prognosis

Aneurysms in the extremities, in contrast with those elsewhere, have an excellent outlook, because sufficient time has elapsed to develop a collateral circulation, and the pathology is readily accessible to surgical therapy.

Treatment

Small aneurysms occasionally are obliterated spontaneously by filling with blood clot followed by fibrous organization. Otherwise, the following surgical measures are indicated:

1. Excision of Aneurysm with End-to-End Anastomosis. This is the procedure of choice when continuity of circulation is vital and it is possible to appose vessel ends by stretching, rerouting, or flexion of adjacent joints. It is especially indicated in young individuals whose vessels are not diseased but whose collateral circulation is inadequate.

2. Excision of Aneurysm With Repair of the Artery. This is applicable especially to traumatic aneurysms. The artery is compressed proximally and distally. The aneurysm is opened widely, and clots are removed. Often the arterial opening is found to be small. The defect is sutured with fine arterial silk, reinforced with a layer of the sac, and the remainder of the sac is excised.

3. Excision With an Analogous Venous or Homologous Arterial Graft. By this method a large gap may be bridged. The saphenous vein is readily available and thickens like the host artery. However, the vein selected should

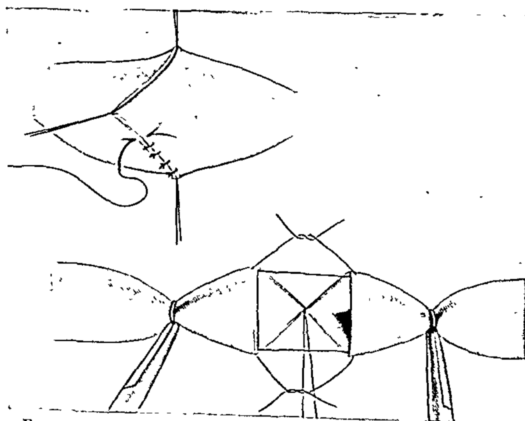


Fig. 258. Triangularization technic. Three sutures are inserted at equidistant points Tension on these sutures makes suturing technically simple. Simple ligation technic illustrated. (Bowers, W. F.: Surgery of Trauma, p. 271, Philadelphia, Lippincott)

plaque often lodges at a point of arterial bifurcation, as at the bifurcation of the aorta, the division into internal and external iliacs, femoral and femoral profunda junction, and division of the popliteal artery. The embolus softens the intimal and the medial layers, which yield to the force of increased arterial pressure that develops behind the obstruction.

C. *Infections.* The arterial wall may be weakened by an infection involving the vasa vasorum, particularly at sites subject to stress and strain, such as the knee. The infection arises from adjacent structures, e.g., actinomycosis (mycotic aneurysm) or, more commonly, from an infective embolus, e.g., subacute bacterial endocarditis. The aneurysm favors an artery unprotected by surrounding muscle and subjected to frequent bending.

D. *Partial Arterial Occlusion.* Poststenotic dilatation occurs typically in the subclavian artery beyond a point of compression. (See Coracobrachial Compression.)

E. *Syphilis* This type of aneurysm results as a late manifestation of untreated syphilis. it occurs predominantly in the aorta, rarely in the extremities, Present-day antibiotic therapy is making the condition extinct.

F. *Arteritis.* Multiple areas of inflammation and necrosis cause countless numbers of small sacculations which are often identified as nodes in periarteritis nodosa.

G. *Less common causes* include burns, roentgen rays, radium, radioactive isotopes, invasion by malignancy, diabetes, lead poisoning, gout and tuberculosis.

Clinical Picture

Symptoms will vary with the vessel involved. Collateral circulation in the upper extremity is excellent, and symptoms of peripheral ischemia are negligible. On the other hand, in the lower extremity claudication is evidence of lesser collaterals.

1. **Tumor Mass.** This may be soft and cystic or somewhat indurated If not obscured by a contained blood clot or by surrounding tissues, it may exhibit expansile pulsations synchronous with systole.

2. **Pain.** This occurs locally due to pressure on adjacent structures, or distally because of nerve compression (neuritic pain) and ischemic claudication.

3. **Pressure Signs.** In addition to pain, nerve compression results in paresis or paralysis; venous compression causes dependent edema.

4. **Signs of Circulatory Insufficiency.** The pulse volume is diminished or absent, oscillometric readings and blood pressure are reduced, and skin temperature is lowered, as compared with the opposite extremity.

5. **Congestion and Edema of the Parts Beyond.** The veins are engorged with blood which flows sluggishly because the arterial pressure is insufficient to move the blood along.

6. **Bruit.** A characteristic sound perceived early on auscultation is due to whirling of blood within the sac. As clotting and fibrosis reduce the size of the cavity, the bruit may disappear. In arterial aneurysms, the bruit is synchronous with systole, in contradistinction to arteriovenous aneurysms, in which there is a to-and-fro murmur.

7. **Trophic Changes.** These occur distal to the aneurysm.

8. **Arteriogram.** This reveals the size and the position of the aneurysm and the extent of collateral circulation.

Pathology

Two types can be identified: traumatic and nontraumatic.

1. **Traumatic Type.** If the wall is partially lacerated, the thin residual wall balloons out and is supported by the surrounding tissue. More often the entire thickness of the wall is divided, and a large hematoma forms within which arterial pressure creates a progressively increasing cavity or sac. Surrounding structures and reactive fibrosis reinforce and fix the walls from without. Laminations of clot are deposited repeatedly on the walls from within and become organized. The thick, leatherlike, endothelial-lined sac may be fusiform or saccular. A true aneurysm microscopically will show the remains of its own thinned-out layers with reactive fibrosis about it. A false aneurysm shows multiple laminations which vary from recent blood clot superficially to older deep layers of fibrous tissue. Aneurysmal pulsations will erode adjacent bone.

2. **Nontraumatic Type.** Ballooning takes place, progressively thinning the wall until rupture occurs, and the surrounding tissues become the sac walls. Laminations of clot organize and thicken the walls. The wall tends

almost invariably by penetrating wounds, is usually single and occurs anywhere in the extremities. The congenital type consists of a tremendous number of communicating channels and is most common about the forearm and the hand. The following discussion is limited to the acquired type of fistula. Congenital arteriovenous fistula is covered in the section on "The Hand."[42]

PATHOLOGY

The fistulous channel is either short or long, and narrow or broad and distended as to constitute an aneurysmal sac. Locally, the vein

artery proximal to the fistula becomes dilated and the walls thinned. Distal to the site of communication, the artery is narrowed. The artery and the vein may enter the fistula or sac separately or through a common opening.

CLINICAL PICTURE

The following are typical:

1. **Profuse but controlled hemorrhage early** after an injury.

2. **Thrill and bruit continuous throughout** the cardiac cycle develops within a few hours or days.

3. **Venous Insufficiency.** Consequent varices, edema, stasis pigmentation, ulceration and chronic indurative cellulitis. These complications involve the extreme distal parts of the extremity in contrast with the chronic venous insufficiency of thrombophlebitis or primary varicose veins which causes stasis ulceration at or above the malleoli of the ankle.

4. **Gangrene** of digits or distal parts of the extremity due to ischemia.

5. **Increase in length of a limb** if fistula is acquired before the epiphyseal lines close.

6. **Increased skin temperature** about and just distal to the fistula.

7. **Cardiac enlargement** due to increased pressure of return blood flow. This occurs only in large caliber communicating channels and may result in congestive heart failure.

[42] Allen, E. V., Barker, N. W., and Hines, E. A.: Peripheral Vascular Diseases, p. 519, Philadelphia, Saunders, 1946.

8. **Increased Oxygenation of Venous Blood.** A blood sample taken from a vein proximal to the fistula is bright red and arterial as compared with a sample from the opposite extremity. Actual measurement of oxygen content reveals the increase.

9. **Branham's Sign.** A sharp decrease in pulse rate occurs when the fistula is closed by digital pressure. The systolic and the diastolic pressures increase at the same time.

ARTERIOGRAPHY

Injection of a contrast media into the artery proximal to the anastomosis, and while the blood flow in the artery is stopped by a tourniquet, reveals the artery to be enlarged and tortuous proximal to the fistula. Distally, the artery is narrowed or not visualized. Locally, at the anastomotic site the vein displays a rounded saccular bulge, and the veins distally are well seen and are enlarged, tortuous and multiple.

OSCILLOMETRY

When the fistual is large and the blood flow easily short-circuited, the pulsations in the distal regions are diminished or absent. When the fistula is small, a compensatory increase in pulse pressure may increase the reading.

TREATMENT

The surgical indications include: (1) chronic venous insufficiency; (2) cardiac enlargement; and (3) prevention of growth increase. A period of at least 6 months waiting is allowed for establishment of collateral circulation. Before repair, the adequacy of collateral [illegible]

Mosz[illegible]

and a t[illegible]

after which the extremity is placed in a horizontal position, and the bandage is removed quickly. When circulation is adequate, a hyperemic blush occurs promptly. When arterial insufficiency exists, the blush is absent or slight and progressing slowly toward the periphery.

If collateral circulation is found to be inadequate a further waiting [illegible]

ally pro[illegible]

may be [illegible] spasm

be comparable in size with the artery. The vein should be so placed that its distal end *joins the proximal end of the artery and vice* versa because of the valvular system in the veins.

Arterial grafts must be taken from young individuals within a few hours after their demise. The grafts can be kept in an electrolyte solution (glucose, serum) containing antibiotics and stored at a temperature of 1° to 4° C. They can be used successfully over a period of 6 weeks. The arterial graft after transplantation acts as a bridge over which a new vessel is formed.

4 **Obliteration of the Aneurysm (Aneurysmorrhaphy of Matas[37]).** This procedure preserves the aneurysmal wall through which collateral vessels have developed. It is the procedure of choice when the above-described procedures are not feasible, particularly in large arteriosclerotic aneurysms. The artery is compressed proximally and distally. The sac is opened, and its contents are evacuated. From within the sac all collateral openings are closed with silk sutures. The main afferent and *efferent vessels are ligated The sac is* retained. A muscle flap placed into and covered by the remains of the sac helps to obliterate the latter [38] It may also be advisable to open the accompanying vein, removing any clots and ligating and dividing the vein.[39] Perhaps this acts to remove the reflexly induced vasoconstriction of the collateral circulation

5. **Perianeurysmal Irritation.** If the aneurysm is obliterated very gradually, collateral circulation will develop to supply adequately parts distal to the aneurysm In the manufacture of a form of polythene, a substance called diacetyl phosphate forms This substance, when placed with a polythene covering[40] around an aneuyrsm, causes the latter to become obliterated by fibroblastic proliferation. A nonirritating cellophane[41] should be placed about the polythene to protect adjacent structures. The polythene should not be tied in place, because the aneurysm will force itself against this irresistible band until it ruptures. *This method is used when other therapy* is impossible, the aneurysm is inaccessible, or the patient's condition precludes further surgery.

Preoperative Management. Before surgery postural exercise tests are conducted for determining adequacy of collateral circulation. Intermittent compression of the vessel by an inflatable tourniquet will encourage the formation of new collaterals. Sympathectomy should precede the operation.

Postoperative Management. The limb must not be elevated and is surrounded by a heat cradle. Papaverine (½ gr.) is administered every 4 hours. The use of anticoagulants is optional.

Aneurysms of the extremities are the interest of the orthopaedic surgeon. An attempt must be made to re-establish continuity of the artery by end-to-end anastomosis, removal of the sac with repair of the opening, a graft of the analogous vein type or homologous artery type, or the obliterative aneurysmorrhaphy of Matas *Gangrene is more likely to follow* complete extirpation of the sac than if the sac wall with its contained collateral circulation is left behind.

Complications of aneurysm surgery in an extremity include (1) hemorrhage, (2) arterial thrombosis, (3) pulmonary embolism, (4) arterial embolism and (5) infection. Later, deficient blood supply is evidenced chiefly by claudication on overuse of the part. The patient must be taught to live within the limits of his circulation until the collateral circulation becomes adequate. Crutches may be required postoperatively. Other signs of ischemia include infections which fail to heal, trophic changes in the digits, and frostbite on exposure to cold. The patient must obstain from smoking; the part must be kept clean and protected from injury; and collateral circulation should be stimulated by warm sitz baths, postural and graded exercises, alcohol and papaverine hydrochloride.

ARTERIOVENOUS FISTULA

An arteriovenous fistula is an abnormal communication between an artery and a vein proximal to the capillary bed. It is congenital or acquired. The acquired type is caused

[37] Matas, R : Endoaneurysmorrhaphy, Ann Surg 27:161, 1903

[38] Pratt, G. H : Surgical treatment of peripheral aneurysm, Surg , Gynec. & Obst. 75 103, 1942.

[39] ———. Surgical treatment of aneurysms, Am. Heart J. 38 43, 1949

[40] Polythene type NV-7-14 (DuPont)

[41] Cellophane type 300 PUT-71 (DuPont).

by sympathectomy after preliminary testing by procaine block.

Technic. An inflatable tourniquet is placed and held in readiness. It is advantageous to have continuous blood flow which permits localization of the abnormal communication by detecting the bruit. The artery is dilated proximally; the vein is dilated both proximally and distally. Then the lowest point at which Branham's bradycardia phenomenon can be elicited is determined. The fistulous area is usually engulfed by scar tissue resulting from the original injury. The affected vessels are ligated above and below the scarred area, and the fistula with adjacent segments of artery and vein is excised. If the collateral circulation is inadequate and the fistulous tract is long, the latter is ligated with heavy black silk, and heparin or Dicumarol is given postoperatively. When the artery is ligated, the accompanying vein must always be ligated, as this allows a more even distribution of blood to the extremity, and there is less likelihood of gangrene. Postoperatively, a loose dressing is applied, the extremity is kept horizontal, and a heat cradle maintains a temperature of 90° F. If the veins are distended and venous congestion is

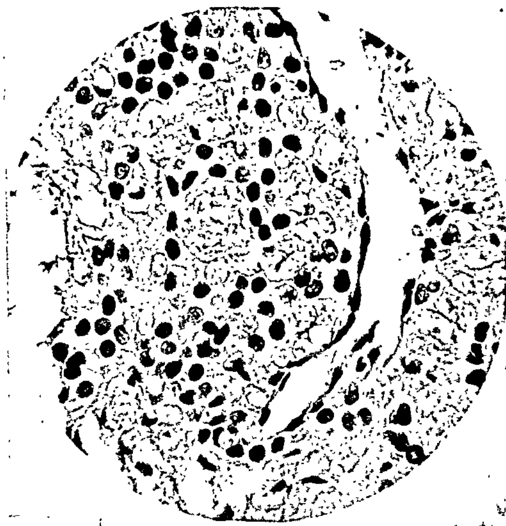


FIG. 259. (*Continued from facing page*)
(D) Higher magnification of microscopic section (× 430). Note the vessel and glomus cells. The blood vessel is lined with endothelial cells, and the surrounding epithelioid cells have large, oval, darkly stained nuclei. The cytoplasm is faintly stained. (Posch, J. L.: Tumors of the hand, J. Bone & Joint Surg 38A:527)

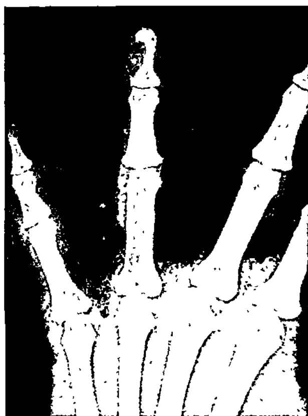




FIG. 259. Glomus tumor. (A, *top, left*) Roentgenogram demonstrating severe erosion of the distal phalanx. (B, *top, right*) Operative appearance at typical site. (C, *bottom*) Photomicrograph (× 90) of encapsulated tumor.

(*Continued on facing page*)

males in the ratio of 2 to 1. No age group is spared, but individuals in the third and the fourth decades are predisposed.

Experimental Work

Polyarteritis can develop in patients during a serum sickness type of anaphylactic reaction.[44] It can be produced experimentally by subjecting animals to serum sickness.[45] The frequency of the condition has increased coincident with the advent and the widespread use of sulfa drugs. Many sensitizing drugs used to reproduce the disease, associated allergic conditions and eosinophilia in many patients suggest but do not prove an allergic origin.

Pathology[46]

The basic lesion is a necrotizing, inflammatory, obliterative process involving the small arteries, the arterioles and occasionally the veins. The following stages delineate the process:

1. **Edema, eosinophilic fibrinoid necrosis** of the inner media and collagenous tissue in the subendothelial and adventitial areas of the arterial wall. Necrosis of media leads to aneurysmal dilatation.
2. **Mononuclear cellular infiltration** of adventitia and media occurring coincident with fibrinoid necrosis In early stages, eosinophils appear in large numbers.
3. **Endothelial destruction leading to thrombosis and infarction**, especially seen in kidneys, intestines, liver, spleen and myocardium. The thrombus becomes organized and eventually recanalized.
4. **Healing stage** of fibroblastic repair of damaged periarterial tissues.

Grossly minute and microscopic nodules along the course of the vessel consist of periarterial inflammation and fibrosis or aneurysm formation. Widespread involvement of organs and tissues is usual.

[44] Rich, A R · Role of hypersensitivity in periarteritis nodosa, Bull Johns Hopkins Hosp 71·123, 1942.

[45] Rich, A R, and Gregory, J. E. Experimental demonstration that periarteritis nodosa is a manifestation of hypersensitivity, Bull Johns Hopkins Hosp. 72 65, 1943.

[46] Arkin, A : A clinical and pathological study of periarteritis nodosa, Am. J. Path. 6:401, 1930



Fig. 260. Periarteritis nodosa. Typical appearance of lesion in biopsied muscle specimen.

Clinical Picture

Symptoms include those of a systemic inflammation plus those specific for the affected regions. The main features are:

1. Fever, weakness, prostration
2. Leukocytosis with a shift to the left; often an eosinophilia
3. Albuminuria and microscopic hematuria
4. Abdominal pain
5. Signs and symptoms of polyneuritis
6. Hypertension

The onset is insidious, with vague symptoms of fever and malaise. At first fever may persist without other symptoms. Later, symptoms appear in various, widely spread, unrelated areas.

Renal involvement consists of albuminuria, microscopic hematuria and cylindruria. Glomerulonephritis, when present, causes generalized edema. Renal insufficiency leads to progressive intractable hypertension.

Myocardial involvement, by fibrinoid degeneration of myocardial collagen, causes abnormalities of cardiac function. Pericarditis and coronary symptoms may occur.

Pulmonary involvement causes diffuse pulmonary infiltrations. Intractable asthma may dominate the picture.

Gastro-intestinal symptoms include persistent abdominal pain, anorexia and vomiting. Hemorrhagic lesions of stomach, bowel, pancreas, liver and spleen produces symptoms referable to these organs.

Polyneuritis symptoms are conspicuous in many cases and include hyperesthesias, numbness, muscle weakness, etc.

present, the extremity is elevated. If ischemia is present, the extremity is lowered. Heparin and Dicumarol prevent thrombosis which might compromise the result.

✓ GLOMUS TUMOR

Throughout the body lying in the stratum reticulare between the skin and the subcutaneous layer are innumerable specialized direct connections between terminal arteries and veins. Each arteriovenous anastomosis or shunt, the glomus or neuromyo-arterial glomus, is under the direct control of the sympathetic nervous system and serves the functions of (1) local and general regulation of heat and (2) the regulation of blood pressure. The structure of the normal glomus was first described by Sucquet and Hoyer.[43] The glomus is an arteriovenous anastomosis with thick tortuous walls. Its canal is lined with several rows of cuboidal endothelial cells beneath which the elastic layer is absent. A thick muscular coat of smooth muscle surrounds the endothelium. Within and around the muscle cells are large epithelioid cells with clear or vacuolar cytoplasm and vesicular nuclei. They constitute the typical "glomus cells" whose origin is unknown. Closely associated with the glomus cells is an abundance of nonmyelinated nerve fibers which are demonstrable by special stains. The tortuous anastomotic channel is designated the "Sucquet-Hoyer canal."

Pathology

The glomus tumor is probably a hypertrophy of the normal glomus. Grossly, the tumor is encapsulated, a few millimeters in diameter, deep red or purple in color, and on cutting exudes blood, following which it assumes a gray color. Microscopically, an overgrowth of the cellular constituents, particularly the epithelioid cells, is noted.

Clinical Picture

Trauma often precedes the tumor. Although glomus tumors may appear anywhere in the body, they are most frequent on the hands and the feet, particularly beneath the fingernails. The glomus tumor typically appears as a small, painful, purplish nodule in the skin or under the nail. A severe neuralgic type of pain may be present for some time before the bluish spot just a few millimeters in diameter appears. The spot or nodule is excruciatingly tender and when pressed upon may become blanched. The lesion can be localized accurately by careful testing with the point of a pin. The normal or surrounding skin is not excessively sensitive, but as soon as the glomus tumor is touched excruciating pain is provoked. The application of cold to the lesion will produce a paroxysm. Occasionally associated with the severe pain are vasomotor changes which may involve the extremity or the entire half of the body. The lesion may occur at any age.

Roentgenologic Findings

The tumor being pulsatile, when it is situated in a finger it may cause a smooth saucer-shaped erosion of the adjacent phalanx.

Treatment

Complete excision will effect a cure. To ensure complete removal, the phalanx should be curetted thoroughly. When using a local anesthetic, infiltration must avoid the vicinity of the tumor where swelling incident to the injection would blanch the tumor and make its identification difficult.

PERIARTERITIS NODOSA (Polyarteritis)

Periarteritis nodosa is a progressive, usually fatal disease of protean manifestations secondary to widespread focal involvement of the walls of medium and small-sized arteries. Fibrinoid degeneration in the walls of blood vessels is characteristic and classifies this as a collagen disease. It can involve any region of the body. Its importance to the orthopaedic surgeon lies in differentiating it from conditions, which it may imitate, in and about the musculoskeletal system. For example, periarteritis in the neck may suggest lesions of the cervical spine; involvement of a plantar digital artery in the foot may be confused with Morton's toe. Eventually, multifocal involvement may clarify the diagnosis, but a correct antemortem interpretation is unusual.

Etiology

The cause is unknown. The affliction favors

[43] Popoff, N. W.: The digital vascular system; with reference to the state of glomus. Arch. Path. 18:295, 1934.

arteritis nodosa), acute arterial obstruction (embolism, thrombosis, injury, surgical division). The aim is to improve the collateral circulation.

3. **Alleviation of Sympathetic Pain:** ischemic pain, causalgic burning pain, and minor causalgic states as in thrombophlebitis, arthritis, etc.

Test of Effectiveness of Sympathetic Interruption

The degree of vasodilatation to be obtained can be evaluated by anesthetic injection of the peripheral nerve, spinal anesthesia, or general anesthesia, but it is best to determine this by anesthetization of the sympathetic trunk, which also relieves pain of sympathetic origin. Vasodilatation can be measured by comparing the rise of digital skin temperature with that in the opposite limb. In obliterative arterial disease, lack of significant alteration is not unusual; however, improvement of symptoms by sympathetic interruption is often obtained. The cessation of sweating and increase in cutaneous electric resistance in a part is proof that the sympathetic trunk has been anesthetized. These tests are useful when no vasodilatation is demonstrable. Autonomic blocking agents, such as tetraethyl ammonium chloride, are of no value because they effect a varying degree of vasodilatation and in effective doses often cause a fall in blood pressure.

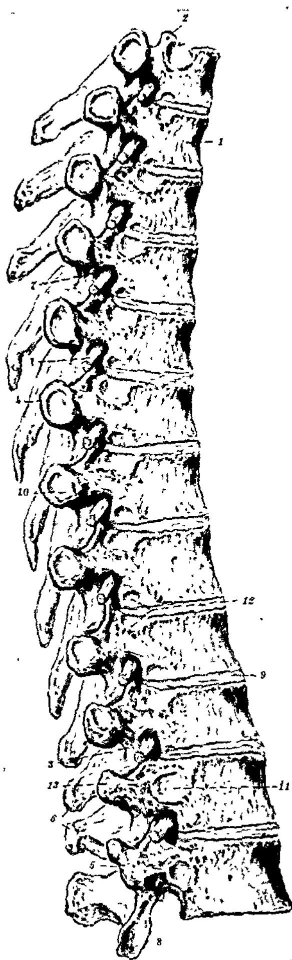


Fig. 261. Thoracic vertebrae as seen from the right side.

Nerves: (1) spinal nerve emerging from the intervertebral foramen.

Tissues: (1) body; (2) superior articular surface; (3) lamina; (4) transverse process; (5) mammillary process; (6) spinous process; (7) intervertebral foramen; (8) inferior articular surface; (9) intervertebral cartilage; (10) facet on transverse process for articular part of tubercle of rib; (11) facet for head of rib; (12) demifacet for articular head of rib, (13) no facet on transverse process. (Southworth, J. L., Hingson, R. A., and Pitkin, W. M. (eds.): Pitkin's Conduction Anesthesia, Philadelphia, Lippincott)

Central nervous system symptoms include convulsions, meningitis, paralysis, etc., as a result of ischemic foci.

Cutaneous manifestations are extremely variable. Occasionally, hemorrhagic lesions may ulcerate.

Muscle and joint pains are characteristic. Although nodules are not palpable, multiple tender spots are identified. Discomfort is persistent and accentuated by active and passive motion. Extensive involvement with multiple foci of ischemic necrosis may materially weaken the muscle.

Cutaneous nerve pain in any specified area is often severe, lancinating and obstinate. It is due to an arteritic process in nutrient vessels of the nerve.

Laboratory Findings. Leukocytosis is moderate, and the sedimentation rate is elevated. Eosinophilia, when present, may be quite high.

Diagnosis

The disease is suspected in a prolonged systemic illness in which several unrelated organs are involved. Biopsy of muscle tissue at a tender site may reveal the characteristic vascular lesion and a sharply circumscribed area of swollen, hyalinized, disintegrating muscle fibers. When a well-localized point of tenderness can be identified beneath the skin proximal to a persistently painful part, one should not hesitate to explore, resect and examine microscopically the cutaneous nerve.

Prognosis and Treatment

Until recently the prognosis was grave; most patients succumbed to renal or cardiac insufficiency or intercurrent infection. A sensitizing antigen should be sought and eliminated. Cortisone and ACTH dramatically suppress and prevent the development of further clinical manifestations. However, if treatment has been started after irreversible damage has occurred in vital organs, the outlook continues to be serious. Some cases become permanently arrested spontaneously and undergo complete healing.

THERAPEUTIC SYMPATHETIC PARALYSIS

Anatomic Facts

The sympathetic portion of the autonomic nervous system consists of two chains of ganglia, each of which lies on the anterolateral aspect of the spine. Each chain is composed of 24 ganglia, 3 cervical (superior, middle, inferior), 12 thoracic, 4 lumbar, and the rest sacral. The white rami communicantes, which contain the efferent fibers from the cord, and the gray rami communicantes, which contain the afferent fibers to the cord, connect the spinal nerve root with its corresponding ganglion in the chain. The cervical ganglia are exceptions. They have no connection with the cervical nerves and instead reach the spinal cord through the upper thoracic nerves, by way of the thoracic ganglia. The upper 3 thoracic ganglia are therefore the sympathetic supply stations for the upper extremity. Occasionally, the inferior cervical ganglion is fused with the first cervical, forming the stellate ganglion. In the thoracic area, the thoracic chain lies in relation to the heads of the ribs In the lumbar area, the chain lies between the medial border of the psoas muscle and the spine. On the right side it is overlapped medially by the vena cava. Also on the right side, the lower lumbar veins pass anterior to the lower segment of the trunk. Elsewhere they pass behind the trunk. The first lumbar ganglion lies at the margin of the diaphragmatic crura. The fourth lumbar ganglion lies just above the sacral promontory.

Indications for Sympathetic Block

The orthopaedic surgeon is concerned mainly with interrupting sympathetic function to the extremities. The sympathetic fibers pass distally through the extremities through the peripheral nerves supplying vasomotor, sudomotor and pilomotor impulses and returning visceral painful impulses to the spinal cord. Therefore, the main uses for sympathetic interruption are reduction of sweating, effecting vasodilatation and relieving sympathetic pain. The following conditions are benefited:

1. **Vasospastic Conditions:** Raynaud's disease, Raynaud's phenomenon as a result of some cause as the use of high frequency vibrating tools, acrocyanosis, frostbite, trench foot or immersion foot, causalgia, livedo *reticularis*, *segmental arterial spasm*, postparalytic cold extremities and thrombophlebitis

2. **Arterial Obstructive Conditions:** chronic obliterative arterial disease (arteriosclerosis *obliterans*, *thromboangiitis obliterans*, peri-

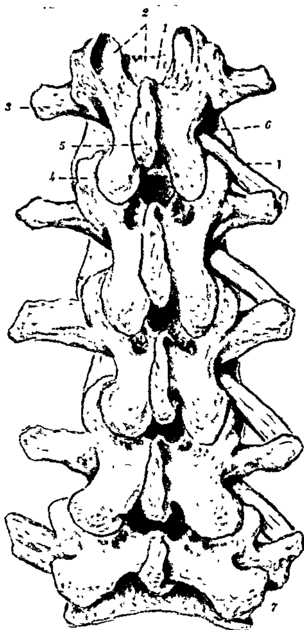


FIG. 264. Lumbar vertebrae, posterior view. Note that in blocking near the midline (1½ inches) as described in the text the needle glides off the inferior border of the transverse process to touch the posterior surface of the body of the vertebra at or near the intervertebral foramen. (See Fig. 265)

Nerves: (1) first lumbar nerve emerging from the intervertebral foramen.

Tissues: (1) body; (2) superior articular surface; (3) transverse process, (4) mammillary process; (5) spinous process; (6) intervertebral foramen; (7) inferior articular surface. (Southworth, J. L., Hingson, R. A , and Pitkin, W. M. (eds.): Pitkin's Conduction Anesthesia, Philadelphia, Lippincott)

TECHNICS OF SYMPATHETIC BLOCKS[47]

Injection of a local anesthetic along the side of the spinal column blocks the nerve roots at their point of emergence from their foramina and the sympathetic chain. This is known as a *paravertebral block*. By blocking one nerve and its corresponding sympathetic ganglion, one visceral and somatic segment of the body is anesthetized (*segmental block*). The technic of paravertebral block is such that it is almost impossible to block the nerve root without similarly affecting its rami to the sympathetic trunk. On the other hand, the sympathetic ganglion may be blocked without affecting the nerve root. The following description pertains to thoracic and lumbar paravertebral blocks, the technic being varied as indicated to obtain sympathetic blocks. For the upper extremity, the lowest cervical and the upper 3 thoracic ganglia are infiltrated. All 4 lumbar ganglia must be blocked to deprive the lower extremity of sympathetic innervation.

Landmarks. In the thoracic spine, the emerging nerve lies at the level of the tip of the spinous process of the vertebra directly above it. For example, the foramen of the first thoracic nerve will be found directly opposite the tip of the spinous process of the 7th cervical vertebra.

In the lumbar spine, the foramen is directly opposite the center of its corresponding spinous process.

The foramen in the thoracic spine lies about 1¼ inches deep to the posterior surface of the transverse process. In other words, when the needle point contacts the posterior surface of the transverse process and then is directed deeply below the inferior margin of the transverse process, it is inserted to a depth of 1¼ inches before it contacts the nerve, the foramen, or the posterolateral aspect of the body of the vertebra. In the lumbar spine, the distance from the transverse process to the foramen is about ¼ inch less.

In the upper thorax, the ganglia lie beneath the necks of the ribs and near the thoracic roots, along the sides of the vertebral bodies. In the lumbar area, the ganglia lie a little

[47] Southworth, J. L., Hingson, R. A., and Pitkin, W M (eds.)· Pitkin's Conduction Anesthesia, ed 2, Philadelphia, Lippincott, 1946

Fig. 262. (*Left*) Posterior view of the thoracic vertebra; spinal nerves emerging from the foramina on right side.

Nerve: (1) spinal.

Tissues: (1) superior articular surface; (2) lamina; (3, 4) transverse processes; (5) spinous process. (Southworth, J. L., Hingson, R. A., and Pitkin, W. M. (eds.): Pitkin's Conduction Anesthesia, Philadelphia, Lippincott)

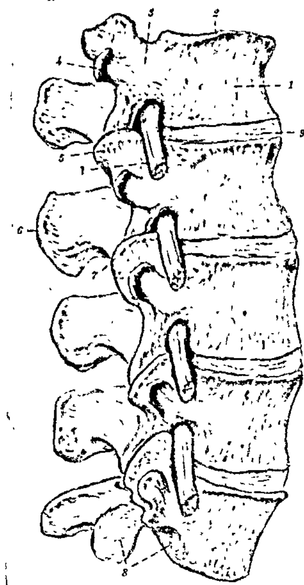


Fig 263. Lumbar vertebrae seen from the right side.

Nerves: (1) first lumbar nerve emerging from the intervertebral foramen.

Tissues: (1) body; (2) superior articular surface, (3) lamina; (4) transverse process; (5) mammillary process; (6) spinous process; (7) intervertebral foramen, (8) inferior articular surface; (9) intervertebral cartilage. (Southworth, J. L., Hingson, R. A., and Pitkin, W. M. (eds.): Pitkin's Conduction Anesthesia, Philadelphia, Lippincott)

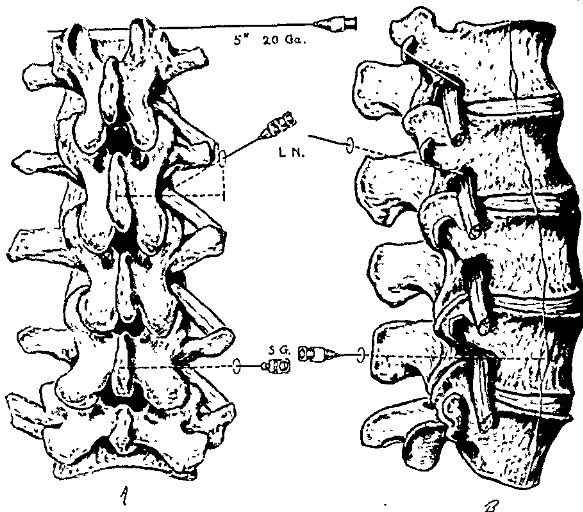
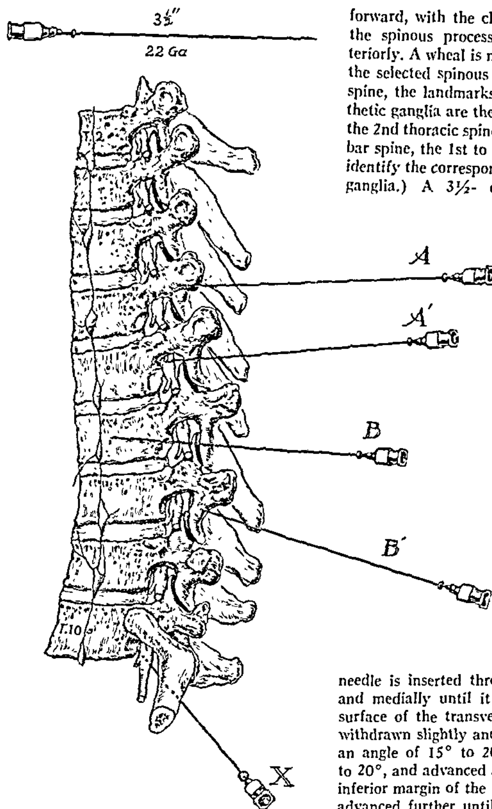


FIG. 266. Technic of paravertebral block in the lumbar region. Upper needle in A shows the correct approach, the needle having glided off the transverse process to the intervertebral foramen (upper needle in B) where 5 cc. of solution will block the somatic nerve and its connections in the upper lumbar region. To block the sympathetic ganglion alone, the needle approach may be made from a site a little more lateral and without downward angulation to by-pass the transverse process until the point comes to rest on the anterolateral aspect of the body of the vertebra. (Southworth, J. L., Hingson, R. A., and Pitkin, W. M. (eds.): Pitkin's Conduction Anesthesia, Philadelphia, Lippincott)

to the sympathetic chain. If the injection of the sympathetics is proper, it will be evidenced clinically by vasodilatation, redness, warmth and dryness of the part. When infiltration of the first thoracic ganglion spreads to involve the inferior cervical ganglion, a Horner's syndrome will be produced.

Stellate Ganglion Block. Occasionally, it is desirable to block the inferior cervical ganglion. This ganglion, when combined with the first thoracic ganglion, is known as the *stellate ganglion*. The inferior cervical ganglion lies immediately in front of the transverse process of the 7th cervical vertebra and in front of the 1st rib and just behind the vertebral artery. It may be approached in the same manner as described for paravertebral block of the 1st thoracic nerve and 1st thoracic ganglion Occasionally, it may be easier, particularly in the obese patient, to approach it from the side.

TECHNIC. A point is selected on the lateral aspect of the neck even with the transverse process of the 6th cervical vertebra, if it is palpable, or a fingerbreadth above the 7th cervical spinous process. Anterior to the trapezius the needle is inserted and directed downward and medially at an angle of about 90° with the mid-line to touch the transverse process of the 7th cervical vertebra. Then the needle is withdrawn slightly and advanced



forward, with the chin on the chest to make the spinous processes more prominent posteriorly. A wheal is made 1½ inches lateral to the selected spinous process. (In the thoracic spine, the landmarks for the upper 3 sympathetic ganglia are the 7th cervical and 1st and the 2nd thoracic spinous processes; in the lumbar spine, the 1st to the 4th spinous processes identify the correspondingly numbered lumbar ganglia.) A 3½- or 4-inch blunt-pointed

Fig. 265. Technics of paravertebral block. (For explanation see text.) (Southworth, J. L., Hingson, R. A, and Pitkin, W. M. (eds.): Pitkin's Conduction Anesthesia, Philadelphia, Lippincott)

needle is inserted through the wheal inward and medially until it contacts the posterior surface of the transverse process. Then it is withdrawn slightly and directed downward at an angle of 15° to 20°, tilted laterally 15° to 20°, and advanced again to pass below the inferior margin of the transverse process It is advanced further until it contacts the posterior surface of the body of the vertebra at the outer border of the intervertebral foramen. Contact with the nerve root may evoke paresthetic sensations. This is the position for injecting the nerve root

If the needle angulation is reduced, that is, the needle hub is moved slightly nearer to the mid-line, the needle may be made to advance further along the side of the vertebral body

deeper along the lateral surfaces of the vertebral bodies. The right sympathetic chain is covered anteriorly by the vena cava, and to the right lies the aorta.

Technic of Needle Insertion. The patient is placed in the lateral recumbent position with knees flexed on the abdomen and head bent

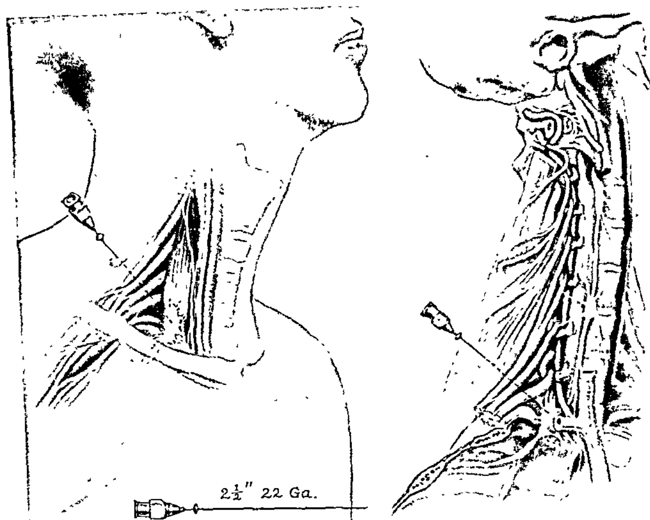


Fig. 268. Lateral approach to the stellate ganglion, indicating the similarity of this technic to that of brachial-plexus block by the lateral route. (Southworth, J. L., Hingson, R. A., and Pitkin, W. M. (eds.): Pitkin's Conduction Anesthesia, Philadelphia, Lippincott)

alongside the body. A pillow is placed beneath the sternum so that the scapulae fall away from the mid-line. An incision is made from the interval between the 1st and the 2nd thoracic spinous processes to the spine of the scapula. The trapezius and the underlying rhomboid muscles are split. This exposes the lowermost fibers of the serratus posterior superior, at the lower margin of which attaches the lumbodorsal fascia. The fascia is incised obliquely, and the serratus is retracted superiorly. Next, the lateral portion of the deep muscles is elevated from the 3rd transverse process and retracted mesially. *It is important to identify accurately the 3rd rib by passing the finger cephalad between the rhomboids and the serratus, locating the uppermost rib and counting downward. The subclavian artery is always palpable above the first rib.* The third rib is exposed subperiosteally, and the transverse process is freed of muscular attachments. An inner segment of rib is removed with the transverse process. The underlying endothoracic fascia must be removed carefully, starting the dissection medially near the vertebral body where the pleura is less intimately associated with the rib than more laterally. By blunt finger dissection, the pleura is pushed away before the rib is excised. Should the pleura be torn, the lung is kept inflated by positive pressure. At the end of the operation the lungs are kept properly inflated as the wound is closed, and no effort is made to close the perforation.

After the rib and the transverse process are removed, the 3rd intercostal nerve is seen lying upon the endothoracic fascia just above the upper margin of the 4th rib. It is picked

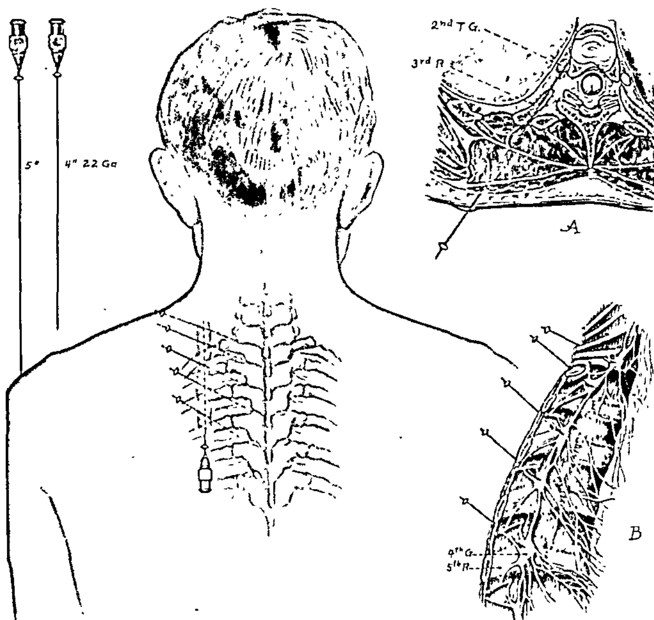


FIG. 267. Thoracic sympathetic ganglion block. (Southworth, J. L., Hingson, R. A., and Pitkin, W. M (eds.): Pitkin's Conduction Anesthesia, Philadelphia, Lippincott)

more anteriorly until it contacts the body of the vertebra, where the injection is made Proper response is indicated by a Horner's syndrome and vasodilatation and dryness of the upper extremity.

The dangers of this method include puncture of the vertebral or subclavian artery, the pleura, or even the dura.

TECHNIC OF SYMPATHECTOMY

Sympathetic Denervation of the Upper Extremity.[48] Removal of, or decentralization of, the upper dorsal ganglia below the level of the first will effect sympathetic paralysis of the upper extremity and half of the head without production of a Horner's syndrome. The operation not only aims at preganglionic denervation of the sympathetics but also measures which avoid postoperative regeneration, namely, removal of the 2nd and the 3rd spinal ganglia and a segment of 2 intercostal nerves, intradural section of the roots, and a silk cylinder to enclose the remaining cut chain ends.[49]

Endotracheal anesthesia is used. The patient lies in the prone position with arms

[48] Shumacker, H B., Jr Sympathetic denervation of the extremities; operative technique, morbidity and mortality, Surgery 24 304, 1948

[49] Smithwick, R H. Modified dorsal sympathectomy for vascular spasm (Raynaud's disease) of the upper extremity, Ann. Surg 104·339, 1936

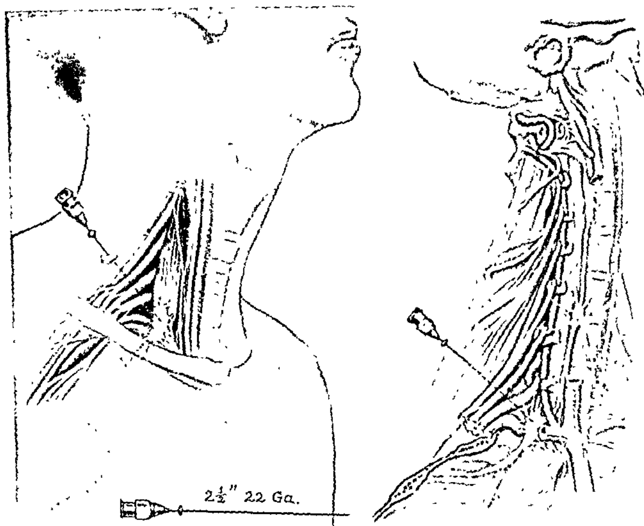


FIG. 268. Lateral approach to the stellate ganglion, indicating the similarity of this technic to that of brachial-plexus block by the lateral route. (Southworth, J. L., Hingson, R. A., and Pitkin, W. M. (eds.): Pitkin's Conduction Anesthesia, Philadelphia, Lippincott)

alongside the body. A pillow is placed beneath the sternum so that the scapulae fall away from the mid-line An incision is made from the interval between the 1st and the 2nd thoracic spinous processes to the spine of the scapula. The trapezius and the underlying rhomboid muscles are split. This exposes the lowermost fibers of the serratus posterior superior, at the lower margin of which attaches the lumbodorsal fascia. The fascia is incised obliquely, and the serratus is retracted superiorly Next, the lateral portion of the deep muscles is elevated from the 3rd transverse process and retracted mesially. *It is important to identify accurately the 3rd rib by passing the finger cephalad between the rhomboids and the serratus, locating the uppermost rib and counting downward. The subclavian artery is always palpable above the first rib.* The third rib is exposed subperiosteally, and the transverse process is freed of muscular attachments. An inner segment of rib is removed with the transverse process. The underlying endothoracic fascia must be removed carefully, starting the dissection medially near the vertebral body where the pleura is less intimately associated with the rib than more laterally. By blunt finger dissection, the pleura is pushed away before the rib is excised. Should the pleura be torn, the lung is kept inflated by positive pressure. At the end of the operation the lungs are kept properly inflated as the wound is closed, and no effort is made to close the perforation.

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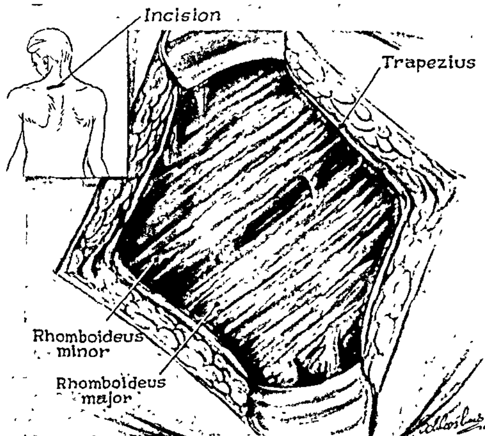




FIG. 269. Dorsal sympathectomy. (*Top*) Showing the position of the patient, the line of incision, and the point of separation of the rhomboids. The trapezius has been split in the direction of its fibers and retracted. (*Bottom*) Showing the incision through the periosteum of the third rib. The lumbodorsal fascia has been incised obliquely below the inferior border of the posterior superior serratus muscle. The deep muscles of the the back are being retracted mesially in order to expose the third transverse process. (Schumacker, H. B., Jr.: Sympathetic denervation of the extremities, Surgery 24: 304)

up, separated from underlying tissues and intercostal vessels, clamped and divided laterally, and traced centrally to the spinal ganglion. The outermost rami communicantes are divided Next, the posterior branch of the spinal ganglion is isolated and divided, thereby *freeing the ganglion and its roots.* The ganglion can be easily delivered thereby so as to expose the remaining rami communicantes and the dorsal and the anterior roots. The remaining rami and the dorsal root are severed Only the anterior root remains connected with the ganglion. By pulling gently on the nerve, the glistening intradural portion of the root is pulled outside the dura. This is divided. Spinal fluid leakage is controlled with a fibrin foam packing.

The 2nd intercostal nerve lies beneath the lower border of the 2nd rib This is freed from its vessels, traced centrally, and the rami communicantes, the dorsal branch and the roots are divided Then the pleura is separated further from the vertebral bodies until the sympathetic chain is isolated, freed and divided between the 3rd and the 4th ganglia The distal end of the proximal portion of the chain is ligated. A silk cylinder is passed over it and ligated gently at its upper end and about the chain above the 2nd ganglion. This ligature must be loose to avoid traumatizing the chain but tight enough to prevent its slipping downward. The distal end of the cylinder is ligated firmly about the end of the freed chain which is then sutured into the adjoining muscles The wound is closed in layers.

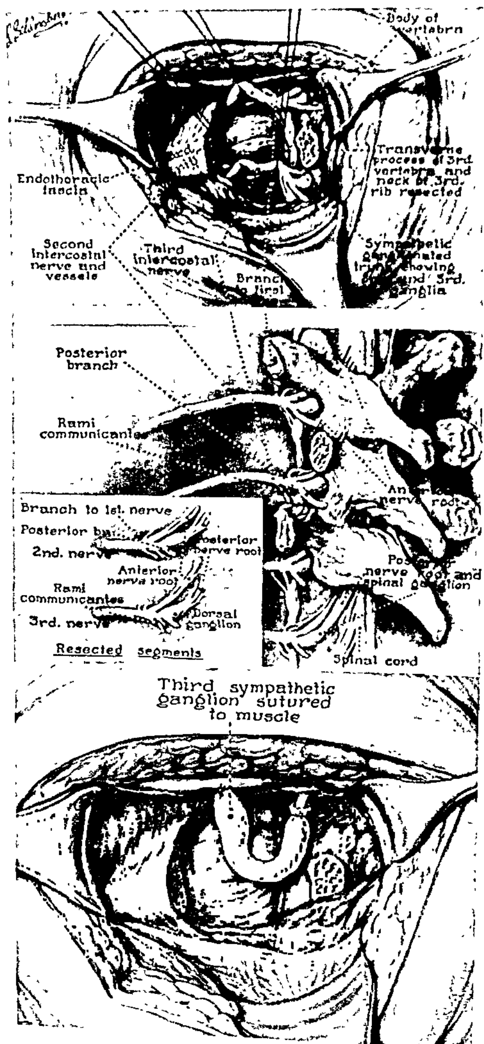


Fig. 270. Dorsal mpathectomy. *'op and center)* ıe third rib and e transverse ocess have been sected. The inter- stal nerves, the rsal ganglia and e sympathetic ain are exposed. tention is called the posterior anches which fix e dorsal ganglia ntrally. (*Bottom*) he decentralized ain has been cov- ed with a silk cyl- der and sutured to ie muscle. (Shu- acker, H. B., Jr.: ympathetic dener- ation of the ex- emities, Surgery 4:304)

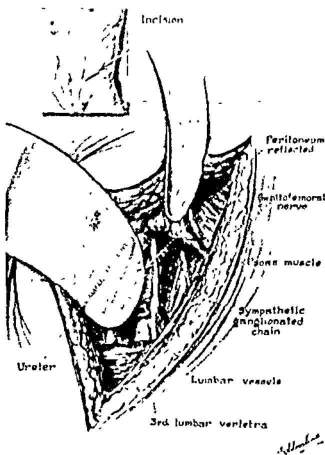


FIG. 271. Lumbar sympathectomy. Anterior muscle-splitting extraperitoneal incision. The retroperitoneal tissues are retracted medially and superiorly to afford exposure as high as the first *lumbar ganglion. (Shumacker, H. B.,* Jr.. Sympathetic denervation of the extremities, Surgery 24·304)

Sympathetic Denervation of the Lower Extremity.[50] Spinal anesthesia provides good muscular relaxation. Curare is administered just before the incision. When sympathectomy is unilateral, the patient is placed on his back, and the side to be operated on is elevated. The lower extremity is placed with the hip flexed to relax the iliopsoas. For bilateral sympathectomy, the patient lies flat on the back with both hips flexed, the table being tilted first to one side and then the other.

A 10-cm. incision extends obliquely from the back at the tip of the 12th rib downward and forward to a point just below and lateral to the umbilicus, following the direction of fibers of the external oblique. The external oblique is split from the mesial to the outer margin of the wound. The internal oblique is exposed at the upper end at its point of fusion with the rectus sheath. It is split from this point downward and posteriorly and bluntly freed from the transversus beneath it. The latter is split and spread far laterally where there is less intimate contact with the peritoneum and less likelihood of tearing the latter. The opening laterally is enlarged by introducing and separating the 2 index fingers, at the same time displacing the peritoneum anteriorly and medially away from the flank and the retroperitoneal tissues. No retractors are used. The dissection must stay close to the peritoneum; otherwise, one may strip off with it a thick layer of retroperitoneal fat and may inadvertently begin dissecting posterior to, rather than anterior to, the iliopsoas muscle. The fingers readily pass over the iliopsoas muscle to the vertebral bodies. Almost invariably at this point one can identify by palpation the sympathetic chain which is felt as a fixed cord of variable size running along the anterolateral aspect of the vertebral bodies.

A broad Dever retractor is introduced in the mesial portion of the wound. The areolar tissue passing from the vena cava or the aorta to the vertebral bodies is incised and *lifted away from the vertebral bodies.* The retractor is reinserted, and its point is held firmly against the vertebral body, thus displacing the vena cava and the aorta mesially and exposing the sympathetic chain. The genitofemoral nerve is seen lying posteriorly upon the iliopsoas muscle. The ureter is visualized as adherent to and elevated with the peritoneum. It contracts when pinched. The lower pole of the kidney is seen and felt in the upper and the outer portion of the wound. The chain is picked up with a long smooth forceps between 2 ganglia, and a loop of silk is passed beneath it for traction. *Elevation of the chain* exposes the rami communicantes, which are divided. Before this is done, the position of the lumbar veins is ascertained. Except for one small vein which crosses the right chain at the level of the 4th lumbar ganglion, the veins usually, but not always, pass posteriorly. *Anterior-lying veins must be ligated and divided.*

The aortic chain of lymph nodes or fascial

[50] Shumacker, H. B.: Sympathetic denervation of the extremities; operative technique, morbidity and mortality, Surgery 24.304, 1948.

bands from the iliopsoas to the vertebral bodies can be mistaken for the sympathetic chain. The chain and the ganglia vary greatly in size but can be identified with certainty by the telltale rami communicantes, the position of the 4th lumbar ganglion just above the promontory, and the position of the first ganglion at the lowermost margin of the diaphragmatic crura at its attachment to the vertebral bodies. The 3rd and the 2nd ganglia vary in size, shape and position and sometimes are fused. The chain is exposed more readily on the left side. On the right side, the vena cava overlies the chain.

The 1st to the 4th ganglia are removed for sympathetic denervation of the lower extremity. In men, the first ganglia are not routinely removed bilaterally unless the patient has first given permission, because of interference with ejaculation. The cut ends of the chain may be left undisturbed.

ANTICOAGULANT THERAPY

Anticoagulant drugs are used to prevent clots and enlargement of clots Their main field of usefulness to the orthopaedic surgeon is in the handling of conditions threatened by thrombotic occlusion of arteries. These substances originated in 1916 when McClean discovered two phosphatides, cuorin and a heparphosphatide,[51] which retarded clotting. Two types of anticoagulants are used, the injectable, rapid-acting ones, of which heparin is mainly used, and oral, slower-acting ones, of which Dicumarol is the best example.

Heparin[52]

This inactivates thromboplastin so that thrombin is unavailable for combining with fibrinogen to form fibrin. It acts rapidly, can be given either intravenously or intramuscularly, and its effectiveness is brief. Three methods of administration are used:

1. 500 cc. of saline containing 10 mg of heparin is given by continuous drip. Coagulation time must be computed every 4 hours.

2. Intravenous injection of 50 to 100 mg. every 4 hours.

[51] McLean, J : Thromboplastic action of cephalin, Am J Physiol 41:250, 1916.

[52] Symposium on Anticoagulant Therapy, Internat M Digest, vol 53, Nov. 1948.

3. Injection in the deep subcutaneous tissues of heparin dissolved in a slowly absorbed menstruum (Depo-heparin*).

Oral drugs are started simultaneously and, when prothrombin time has been increased sufficiently, heparin is discontinued.

Hemorrhage during heparinization can be controlled by giving 50 to 150 mg. of protamine sulfate or toluidine blue, 2 mg. per Kg. of body weight, intravenously. Blood transfusions may be indicated.

Dicumarol[52]

The active substance, dicumarin, is obtained from sweet clover. It acts to increase the prothrombin time. Liver and renal damage are contraindications to its use. When an immediate anticoagulant effect is desired, heparin is given simultaneously with Dicumarol. When the prothrombin time has been raised, only Dicumarol is continued. The latter may be given over any period of time provided that regular prothrombin time determinations are made. A desirable prothrombin time is about 27 seconds, which is equivalent to a prothrombin level of 30 per cent. A rise of prothrombin time over 35 seconds demands temporary stoppage of the drug. A suggested dosage of Dicumarol is 300 mg. the first day, 200 mg. the second day, and 100 mg. per day thereafter Individuals vary in their susceptibility, and the correct dose is that which will keep the prothrombin time at the desired level.

Vitamin K is an antagonist to Dicumarol. In the event of hemorrhage, 60 mg. of synthetic vitamin K (menadione bisulfite) is given intravenously. Whole, fresh blood should be transfused.

Contraindications to Anticoagulant Therapy

1. *Vitamin C and K deficiency and hepatic disease* (anticoagulants unnecessary)

2. *Renal insufficiency*—retention enhances the effect by an abnormal accumulation.

3. *Blood dyscrasias* which impair the clotting mechanism.

4. *Recent brain and spinal cord operations* —hemorrhage is disastrous.

5. *Ulcerative lesions or open wounds* in

* Upjohn Co , Kalamazoo, Mich.

which a tendency to bleeding exists. However, in surgical repair of arterial wounds heparin *is indispensable and may be continued for* about 3 days or until the danger of clotting at the suture site has passed.

DIFFERENTIAL DIAGNOSIS OF ARTERIAL OBSTRUCTION IN AN EXTREMITY

1. Occlusive Arterial Diseases

A. *Arteriosclerosis Obliterans.* The onset usually occurs after the age of 40. Males are affected more commonly but not exclusively The upper extremities are rarely involved (40% in thromboangiitis obliterans); thrombophlebitis is never associated. The arteries are calcified, and hypertension and diabetes are often associated. In younger patients the plasma lipoids are frequently elevated.

B. *Simple Arterial Thrombosis.* This condition *is rare. It is often a complication of severe* infectious disease, blood dyscrasia, congestive heart failure, or trauma. The first manifestation is often an acute, extensive arterial *occlusion with a large area of gangrene*

C. *Arterial Embolism.* This is a complication of bacterial endocarditis, auricular fibrillation, or myocardial infarction with a mural thrombosis. *It* is characterized by sudden arterial occlusion

D *Compression at the Cervicobrachial Junction.* This involves one or both hands. The pulse is reduced or obliterated by various movements of the arms or the head Peripheral nerve involvement is frequent, and cervical rib is often present.

2. Other Vascular Conditions

A. *Raynaud's Disease.* This condition is rare in men. The upper extremities are involved more often Color changes are bilateral and symmetric. Pulsations are present, and mass gangrene of digits does not occur.

B. *Acroscleroderma (Acrosclerosis).* This condition may produce gangrene, rarely occlusion of large arteries. The upper extremities are involved more extensively than the lower. It is bilateral and symmetric. Sclerodactylia is present. (No such skin changes appear in Buerger's disease or arteriosclerosis obliterans.)

C. *Livedo Reticularis.* In this condition arterial occlusion and gangrene rarely occur. The skin is extensively involved, with a livid, reticulated mottling.

D. *Erythermalgia (Erythromelalgia).* The affected parts of the extremity are very warm during painful episodes. Pulsations are normal.

E. *Pernio.* The *affected part is* hot and swollen. Pulsations are normal. The condition develops after exposure to cold. The ulceration is small in extent and situated on the legs *rather than the toes and the feet.*

F. *Ergotism.* This type of arterial occlusion is rare, with a history of medication or food containing ergot. The lesions are symmetric, and often the eyes are involved.

G. *Acute Thrombophlebitis of Large Veins.* The extremity is swollen, the veins are distended, the pulsations may be absent temporarily but soon return, and there are tenderness, redness and pain along the course of a thickened cordlike vein. If pulsations are persistently absent, thromboangiitis obliterans is invariably an associated disease.

H. *Venous Insufficiency.* This condition is due to varicose veins or thrombophlebitis. A congestive pain develops while standing, more so than when walking (in contrast with arterial occlusive disease, which is worse when walking) and is relieved by recumbency. Arterial pulsations are normal.

PART THREE

Regional Orthopaedic Conditions

19

The Cervical Spine →

CERVICAL DISK

Persistent pain in the neck and the upper extremity is usually caused by cervical nerve root compression at the intervertebral foramen. Semmes and Murphey, in 1943, wrote their classic paper which formulated the basic concepts of this condition.[1]

Anatomy

Eight pairs of nerve roots arise from the cervical spinal cord. The first root leaves the spinal canal between the occiput and the 1st cervical vertebra; and the 8th root has an exit between the 7th cervical and 1st thoracic vertebrae. Therefore, a disk lesion between the 6th and the 7th vertebra (6th cervical disk) causes pressure on the 7th cervical root. The dentate ligaments anchor the cervical spinal cord very firmly, and the nerve root runs a short and direct course transversely to the intervertebral foramen. Therefore, the cervical nerve structures are relatively immobile as compared with the remainder of the spinal cord and nerve roots. A cervical nerve root directly overlies the disk space and is immediately subject to pressure when the disk ruptures The cervical disk herniation is very small and usually no larger than a pea However, this small protruding tissue causes enormous pressure because of the limited capacity of the spinal canal and the immobility of the nerve structure.

The lower cervical spine is unusual in that it has 2 pairs of synovial-lined joints surrounding the intervertebral foramen. The posterior border of the foramen is formed by the articulating pedicles of adjacent vertebrae, and the anterior joints of Luschka form the anterior wall of the canal. A degenerative arthritis of these joints may produce spurs that in turn compress the nerve roots and narrow the intervertebral foramen.

Rupture of the cervical disk characteristically occurs lateralward where the annulus fibrosus is weakest and where the posterior longitudinal ligament is thinned. Thus is explained the initial symptoms of unilateral nerve root compression with radicular pain and paresthesia corresponding to the root dermatome.

Pathologic Anatomy.[2] Two types of lesions involve the lower cervical intervertebral foramen and produce identical symptoms. The first type is osteophyte formation encroaching on the intervertebral foramen and associated with narrowing of the corresponding intervertebral disk. This may constitute a late degenerative change resulting from a pre-existent disk lesion with subsequent instability of the joints. These localized arthritic changes characteristically occur at the site of maximum mobility of the cervical spine (5th and 6th cervical junction) and indicate a very probable traumatic etiology. The second type is an acute posterolateral rupture of a cervical disk with immediate compression of the nerve root in the foramen. Neither spur formation nor disk narrowing is demonstrable by oblique x-ray studies Ninety-five per cent of disk lesions occur at the 5th and the 6th levels. The remainder occur at the 4th and the 7th levels.

Etiology

Most frequently, a history of sudden forcible flexion of the neck, the so-called "whiplash" injury, is obtained. The trauma often

[1] Semmes, R. E, and Murphey, F.. Syndrome of unilateral rupture of the sixth cervical intervertebral disc, [illegible] repor[illegible] diseas[illegible]

[2] Keyes, D C, and Compere, E L.: Normal and pathological physiology of nucleus pulposus of intervertebral disc; anatomical, clinical and experimental study, J. Bone & Joint Surg 14:897, 1932.

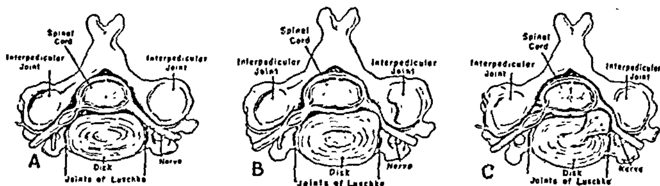


FIG. 272. (A) Normal relationship between the intervertebral disk, the bony structures and the neural elements in the lower cervical region. (B) Mechanism of compression of one of the cervical nerve roots by osteophytic spurs. (C) Lateral rupture of a cervical intervertebral disk with herniation of the nucleus pulposus. (Spurling, R. G., and Segerberg, L. H : Intervertebral disk lesions in the lower cervical region, J.A.M.A. 151:354)

occurs when the individual is sitting in a car which is struck from the rear. A downward compression or hyperflexion mechanism likewise takes place when the head strikes the roof of the car or in a diving accident

SYMPTOMS AND CLINICAL FINDINGS[3]

The discomfort in the neck characteristically occurs early and frequently immediately following the injury. It may be described as a "crick" in the neck or a sense of tightness; or the onset of symptoms may be insidious Exacerbations and remissions are the rule.

[3] Spurling, R. G., and Segerberg, L. H.: Lateral intervertebral disk lesions in the lower cervical region, J.A.M.A 151:354-359, 1953.

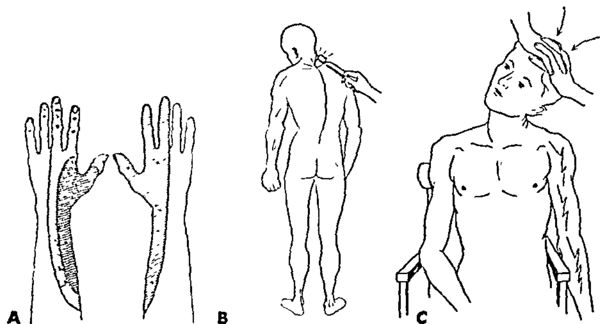


FIG. 273. (*A, left*) Dorsal view of forearm and hand, illustrating the approximate areas into which pain or paresthesias radiate or in which sensation is diminished when the 6th and the 7th cervical roots are compressed. The crosshatched area corresponds roughly to the 6th cervical dermatome, the lined area to the 7th cervical dermatome. (*A, right*) Ventral view of the forearm and the hand illustrates the approximate area into which pain or paresthesias radiate or in which sensation is diminished when the 7th cervical root is compressed. (*B*) Frequently radiating pain can be produced by percussing directly over the lesion. When positive, this test causes pain or paresthesias to radiate into the shoulder and arm (*C*) Method of performing neck compression test in patients with suspected ruptured cervical disks. (Spurling, R. G., and Scoville, W. B.: Lateral rupture of the cervical intervertebral discs, Surg, Gynec. & Obst. 78:350)

The subsequent attacks are usually more severe than the preceding ones and last much longer.

Subjectively, the patient experiences pain in the lower part of the neck radiating into one shoulder and arm and anteriorly and posteriorly over the upper chest area. Paresthesias, such as numbness and tingling, are common over the radial aspect of the forearm, the hand and the fingers. The pain is accentuated by movement of the neck and by coughing, sneezing and straining. The discomfort is severe at night and interferes with sleep. Relief is often obtained by the upright position and ambulation. Movement of the head either forward or backward and usually toward the side of the pain increases the discomfort. The patient usually finds one position of the head, often in neutral flexion and deviation away from the side of the lesion, to gain relief.

Clinically, the following are the findings: marked muscle spasm and limitation of active and passive motion, particularly toward the side of the lesion. The cervical lordosis is reduced. Forcible movement of the head reproduces the pain. Pressure on top of the head likewise accentuates the symptoms. Localized tenderness is felt over the involved disk space slightly to one side of the mid-line. Hypesthesia exists in the 6th or the 7th cervical dermatome. Weakness or fibrillations of the muscles supplied by the 6th or the 7th cervical nerve is sometimes observed. The biceps reflex is reduced in 6th cervical root involvement, and the triceps reflex in the 7th.

Rupture of 5th Cervical Disk: Typical findings:

1. The neck compression test reproduces the pain and causes paresthesias in the thumb and the radial side of the hand.
2. Hypesthesia in the 6th cervical dermatome on the dorsal and the lateral aspects of the thumb and the radial side of the hand.
3. Weakness, atrophy, or fibrillations in the biceps
4. Reduction of the biceps reflex

Rupture of the 6th Cervical Disk:

1. Neck compression test reproduces the pain and causes paresthesias in the index and the middle fingers and the dorsum of the hand.
2. Hypesthesia in the 7th cervical dermatome, the index and the middle fingers and the dorsum of the hand

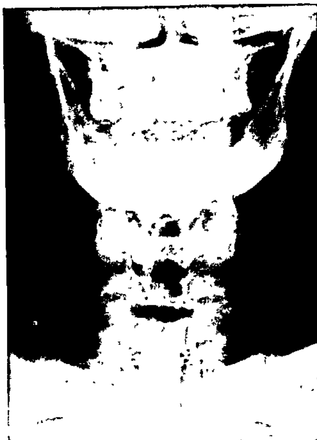


FIG. 274. Flow of Pantopaque is blocked by disk protrusion between the 6th and the 7th cervical vertebrae.

3. Weakness, atrophy or fibrillations in the triceps
4. Reduction of the triceps reflex

Roentgenologic Examination

Views should be taken in the following positions: anteroposterior, lateral and both left and right oblique. The lateral view should be done with the neck in flexion and extension, thereby revealing instability by angulation at one of the cervical junctures. The formation of osteophytic spurs and a narrowing of the foramen is determined on the oblique view. This is generally associated with narrowing of the disk space. Generally, soft-tissue rupture is not associated with disk-space narrowing.

Myelography localizes the lesion preliminary to surgical exploration. The Pantopaque is introduced into the lumbar space with the patient on the fluoroscopic table. The head portion of the table is tilted downward so that the Pantopaque runs over the dorsal hump

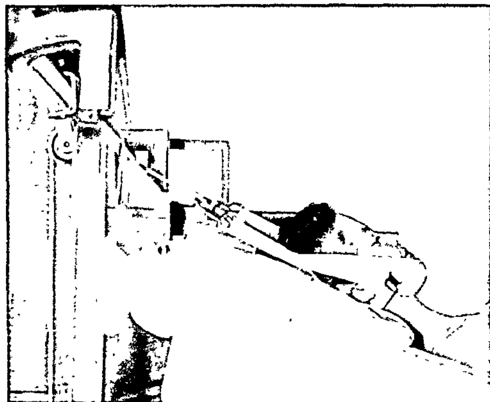


Fig. 275. Head-halter traction. Slight forward-flexion position causes maximum pull through axis at cervical spine.

into the cervical gutter. During this maneuver the head must be held in acute extension to prevent the contrast medium from entering the basal cisternae. Then the table is returned to the horizontal, the cervical spine is studied under the fluoroscope, and films are taken.

The characteristic findings are indentation of the column of Pantopaque or obliteration of the shadow of the nerve-root sleeve at the site of compression. When no indentation is found, but the shadow of the sleeve is absent, the presence of a "hard disk" is probable.

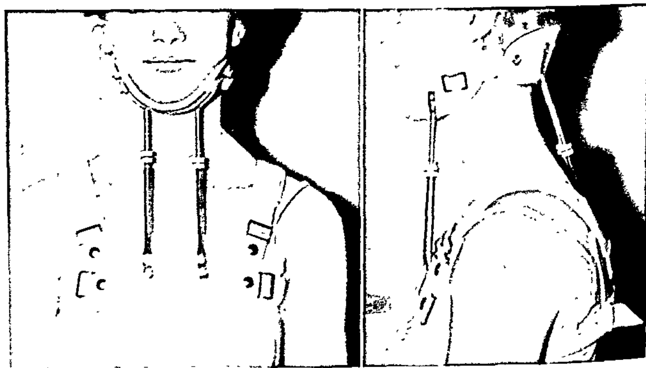


Fig. 276. Cervical brace. The chin and the occipital rests can be raised and distraction of cervical segments effected.

Fig. 277. (*Left*) Posterior view of patient in the upright position for lower cervical hemilaminectomy. Note line of operative incision. (*Right*) Oblique view of patient in the upright position, using the head rest devised by Dr. Winchell McK. Craig, Mayo Clinic, Rochester, Minn. (Spurling, R. G., and Scoville, W. B.: Lateral rupture of the cervical intervertebral disks, Surg., Gynec. & Obst. 78:350)

Differential Diagnosis

The inflammatory lesions about the shoulder and fibrositic nodules about the scapula can produce similar symptoms. Infiltration of the suspected area with procaine relieves the pain and clarifies the diagnosis. The scalenus anticus syndrome is usually characterized by tenderness over the scalenus muscle, sensory diminution in the ulnar distribution, and reduction in volume of the radial pulse. Spurling believes that scalenus compression and its symptoms are secondary to acute lesions in the shoulder or the neck and is comparable with muscle spasm in the lower back with its attendant sciatica. These primary lesions should be sought and ruled out before a diagnosis of primary scalenus anticus compression is made. Cervical cord tumors are ruled out by myelography.

Treatment

Conservative. The majority of cases secure satisfactory relief by conservative treatment. This includes cervical head halter or skeletal traction, complete bed rest, sedatives, hot packs applied at frequent intervals about the neck to reduce muscle spasm, and possibly administration of mephenesin (Tolserol). The patient should persist at this treatment for a long period of time and may rig up a traction apparatus at home. After these initial acute symptoms have subsided, a soft felt collar is applied and is worn continuously to keep neck movement at a minimum. A cervical plaster cast or brace affords the maximum amount of immobilization and occasionally may be indicated. Surgery is done only for persistent or frequently recurring pain and disability.

Operative Technic. The following procedure is described by Kristoff and Odom:[4]

The patient is placed in a sitting position, in

[4] Kristoff, F. C., and Odom, G. L.: Ruptured intervertebral disc in the cervical region, Arch. Surg. 54: 287, 1947.

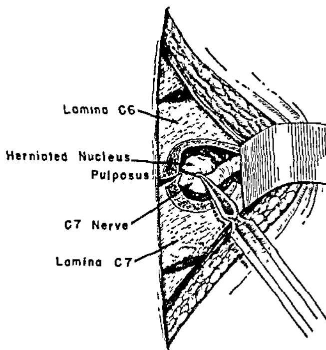


Fig. 278. Diagrammatic sketch of the operative field after the 7th cervical nerve root has been thoroughly decompressed and retracted downward to expose a lateral ruptured disk. (Spurling, R. G., and Segerberg, L. H.: Intervertebral disk lesions in the lower cervical region, J.A.M.A. 151:354)



Fig. 279. Arnold-Chiari deformity. Associated with a thoracic myelomeningocele. Ventral view of spinal cord. Note upward course of cervical nerve roots, diplomyelia and associated midline bony anomaly. (Campbell, J. B.: Congenital anomalies of the neural axis, Am. J. Surg. 75: 231)

the Craig head rest, with the head flexed forward This position not only decreases the bleeding but aids greatly in the exposure of the lesion (Some prefer the horizontal position because of the fear of air embolism in the upright position) Endotracheal anesthesia is used Local anesthesia may be used and is helpful in locating the correct interspace by application of pressure over the ligamentum flavum However, this is extremely uncomfortable for the patient A midline incision is made from the spinous process of the fourth cervical to the first dorsal spinous process, and the posterior muscles are separated in the midline by blunt dissection. The laminae of the fifth, sixth and seventh cervical vertebrae are denuded X-rays should be studied for characteristics of the spinous processes in order that the affected vertebrae be identified Usually a pronounced bifurcation of the tip of the fifth cervical spine and sometimes also of the sixth is found but that of the sixth is always shallower. The seventh cervical, the longest and most prominent, ends in a smooth rounded nodule. After exposure of the laminae the attachment of the ligamentum flavum is separated from the anterior surface of the lamina above The caudal one-third of this lamina is removed and the removal is carried well laterally. In order to accomplish the lateral removal, it may be necessary to use a small dental chisel The ligamentum flavum, which is much thinner than that in the lumbar region, is removed by sharp dissection In most cases this will give an adequate exposure for exploring the interspace, but frequently it is necessary to

remove a small portion of the upper lateral part of the lower lamina. If a disc is present, the nerve root is flattened, displaced posteriorly and exceedingly tense. The root is retracted upward and the herniated disc is removed with forceps. No attempt is made to curet the intervertebral space. Every effort should be made to avoid the large extradural veins, and careful hemostasis should be obtained.

The soft pulpy disk is easy to remove. However, the removal of the hard calcified spur which displaces the nerve root backward is technically difficult. In such cases decompression by removing the posterior wall of the foramen will usually give satisfactory results.

Postoperatively, ambulation is permitted early. The arm pain is nearly always improved or relieved within 24 hours. Numbness and paresthesia may persist for several weeks, and tendon reflex activity may never return to normal. Return to work is permitted in 2 to 4 weeks.

COMPRESSION OF THE SPINAL CORD BY A RUPTURED CERVICAL DISK.[5] Protrusion of disk material at the mid-line is uncommon, but when it does occur will compress the spinal cord and cause symptoms mainly referable to the lower extremities. The herniated disk presses on the anterior aspect of the cord, forces the cord backward and thereby produces traction upon the dentate ligaments, which in turn places tension upon the posterolateral columns—the pyramidal tracts and spinocerebellar tracts. Resultant symptoms include spasticity and hyperreflexia in the lower extremities and unsteadiness of gait. Pain, tenderness and stiffness in the neck are uncommon. The upper extremities present lesser symptoms, including paresthesias, muscle weakness and clumsiness.

When the protrusion is situated slightly more laterally, a Brown-Sequard syndrome results. The area of sensory deficit is usually much lower than the level of the lesion.

Spinal fluid examination in most instances will be negative. Rarely will the Queckenstedt reveal an obstruction of the spinal canal. Spinal fluid protein is normal or slightly elevated, and xanthochromia is infrequent. Pantopaque myelography will reveal deformity in many but not all patients.

Treatment consists of removal of the disk. This is accomplished best through a transdural approach after wide laminectomy of at least two vertebrae. In early cases, the results are *good*.

SIMULATION OF HERNIATED CERVICAL DISK BY THE ARNOLD-CHIARI DEFORMITY.[6] The Arnold-Chiari deformity is characterized by distal displacement of the spinal cord in its relation to the spine. The longitudinal growth of the spine does not keep pace with that of the spinal cord. In consequence, the distal end of the cord, the conus medullaris, extends to the lower lumbar or sacral level where it is held by the shortened filum terminale. The cerebellum, the 4th ventricle and the medulla are pulled distally to protrude through the narrowed foramen magnum, causing a picture of headaches and spastic paresis. Instead of the cervical nerve roots taking a direct lateral course, they are reversed and run obliquely craniad to reach their foramina; on emerging from the foramina, they turn downward toward the upper extremity. Therefore, each root has a double angulation, which may subject it to compression and cause peripheral nerve root symptoms identical with those caused by cervical disk pressure.

The composite clinical picture of Arnold-Chiari deformity and cervical nerve root compression is characterized by headache; pain, paresthesia, and hypesthesia corresponding to the nerve root; difficult urination; muscular weakness and hyperactive reflexes in the lower extremities. Roentgenograms may reveal platybasia and upward invagination of the atlas. Myelography may show a defect similar to that caused by a disk protrusion. Exploratory laminectomy fails to reveal the disk, but the nerve root is seen to take an upward direction. Treatment consists of adequate subtotal occipital craniectomy and opening of the dura. An alternative procedure is sectioning of the filum terminale which permits the nerve structures to recede upward.

Arnold-Chiari deformity should be suspected when cervical disk symptoms arise in the absence of trauma. Conditions to be differentiated are: cord tumor, cervical disk, spur of hypertrophic arthritis, and chronic hypertrophic spinal pachymeningitis. The last-

[5] Bucy, P C, Heimburger, R. F, and Oberhill, H. R.: Compression of cervical spinal cord by herniated intervertebral discs, J Neurosurg 5:471, 1948.

[6] Colclough, J A Surgery 28 874, 1950.

named condition is characterized by bilateral radicular pain, history of an infectious process, and acute percussion tenderness.

DEGENERATIVE JOINT DISEASE OF THE CERVICAL SPINE

The physiologic degenerative aging process, as elsewhere, occurs in the cartilaginous and ligamentous structures of the cervical spine. The process is accentuated by the repeated stresses and strains imposed by this very mobile structure Anteriorly, the disks degenerate, the disk spaces narrow, and marginal spurs develop. The subchondral bone thickens as a result of the increased pressures due to loss of disk substance. The spine angulates forward as the cervical lordosis is reduced. Degeneration of the annular ligament invites rupture and extrusion of disk material. This occurs most commonly at the site of maximal stress, i.e., below the 5th or 6th cervical vertebra.

Degenerative changes after loss of disk substance takes place gradually over many months and are maximal in degree about the involved intervertebral disk space. At the outset, the disk emerges posteriorly to one side or the other, where the restraining ligament is thinner, and compresses the nerve root, causing unilateral radicular symptoms and findings. Later, the disk material is absorbed, the tear heals, and a bony spur develops from the margin of the body and extends backward into the foramen, where it gradually compresses the nerve root and produces a picture of gradually increasing disability. Posteriorly, the degeneration about the facets is maximal at the levels opposite the sites of disk loss and spur formation.

In the lowest cervical vertebrae, additional articulating facets, the joints of Luschka, which lie anterior to the foramina, participate in the degenerative disease. A cervical spine which displays degenerative changes, even though severe, is not necessarily productive of symptoms. The joints which are less involved compensate so that motion is preserved to a remarkable degree. However, some restriction of motion and loss of flexibility is present. A degenerated cervical spine which has restricted motion is predisposed to severe hyperextension injury. Instead of the stress being diffusely distributed throughout the spine, it is thrown upon one site, the posterior laminal elements compressing the cord and causing a paraplegia.[7] Dislocations and fractures are followed by localized degenerative changes because of associated disk damage, instability and incongruities of joint surfaces.

Clinical Picture

Generalized cervical degenerative disease may produce no symptoms and findings, although the condition is obvious in the roentgenograms. On the other hand, the degenerative process in its early stages may not be visible, yet it may cause symptoms. An inflammatory reaction of the soft tissue about the articulating facets is necessary for production of symptoms. Usually, one severe trauma such as suddenly turning the head about or looking upward, or repeated small traumata such as an occupation requiring constant cervical movements, is necessary to cause irritation about the irregular facets and reactive inflammatory synovitis.

Local Symptoms and Findings

Continuous aching *pain* becomes worse with activity and in assuming the erect position; it is relieved by recumbency, heat, salicylates and rest, which reduces cervical motion. *Stiffness* of the neck after rest is relieved by activity. Pain is accentuated with lowered barometric pressures. *Muscle spasm* of all cervical muscles. *Reduction of cervical lordosis* is due in part to the degenerative disease (permanent) and in part to muscle spasm (temporary). A sense of *weakness and inability to hold the head erect* for more than short periods is characteristic. *Tenderness* is acute and well localized laterally over the inflamed articulation. *Cervical motion is limited* in all directions except possibly rotation.

Peripheral Symptoms and Findings

These depend on the particular nerve roots compressed by bony spurs (onset is insidious and progressive, with no remission) or irritated by synovitis of the facet articulation (acute onset, which subsides with remission of arthritis). Symptoms vary from occipital pain (upper cervical), upper ex-

[7] Taylor, A R · The mechanism of injury to the spinal cord in the neck without damage to the vertebral column, J. Bone & Joint Surg. 33B:543, 1951.



Fig. 280. (*Top*) Degenerative arthritic changes in the cervical spine following loss of disk material, particularly between C5 and C6. However, the oblique view shows spur formation encroaching on the intervertebral foraminae from the 3rd to the 7th cervicals, one or all of which can cause nerve root symptoms.

Fig. 281. (*Left*) Fusion of cervical spine from C4 to C6 for pain and instability due to old fracture dislocation of C5.

tremity pains, paresthesias, weakness, altered reflexes and sensory disturbances (middle cervical) to anterior thoracic pain simulating angina pectoris (lower cervical). The most common complaints are referable to the 5th or the 6th cervical disk spaces which involve the 6th and the 7th cervical nerve roots. The neurologic identification is covered under the subject "Cervical Disk" (q.v.).

Roentgenologic Findings

Early, when cartilage alone is degenerating, the roentgenologic picture is negative. Later, the intervertebral spaces narrow, the opposed vertebral body cortices are thickened, dense and irregular. Spurs project from the margins and occasionally may meet, fuse and bridge the gap. Posteriorly, the facets show sclerosis and irregularity and may override one another. The joint space is narrowed. Oblique views are necessary to reveal spurs which project into the intervertebral foramina. Cervical



FIG. 282. Cast for immobilization of the cervical and the upper thoracic spine.

lordosis is reduced. During a period of acute inflammation, the muscle spasm straightens the spine even further. Degenerative changes are more severe at the site of previous disk rupture.

DIFFERENTIAL DIAGNOSIS

Symptoms from degenerative arthritis must be differentiated from cervical ribs, ruptured cervical disk, rheumatoid spondylitis, cervical fracture or dislocation, peripheral neuritis due to other causes (alcohol, avitaminosis, etc.), scalenus anticus syndrome, ordinary sprains and fibrositis.

TREATMENT

Conservative

REST is obtained by *recumbency* and placing *a small pillow under the lordotic curve* to support the cervical spine.

TRACTION. A head halter to which 5 to 15 pounds of continuous traction is applied will distract the irregular facets and reduce the synovitis and the reflex muscle spasm. Traction should be *tried in various lines of pull* until the position of maximum comfort is secured. Usually this is in slight forward flexion. Recently, periods of intermittent traction have been recommended.

MANIPULATION. Gentle passive rotation of the head in various directions while traction is being exerted *is helpful in stretching the* periarticular soft-tissue structures.

HEAT. Moist heat in the form of hot packs is applied. Hot towels are placed about the neck, covered with plastic or waxed paper, then encircled with an electric heating pad Diathermy should be avoided, as it causes bone necrosis and theoretically contributes to further degeneration.

MASSAGE. Massage of muscles after application of heat helps to prevent muscle atrophy and weakness.

SALICYLATES are specific for analgesia. An antiacid is given concomitantly to prevent gastric irritation.

CORTISONE AND ACTH act by their anti-inflammatory action but are not as effective as in rheumatoid arthritis. Their use is not recommended, particularly in view of possible adverse side effects.

LOCAL COUNTERIRRITATION by methyl salicylate is supposed to be beneficial by producing hyperemia. This author believes the effect *is purely psychologic.*

ORTHOPAEDIC APPLIANCES. Restriction of cervical motion permits subsidence of inflammation. A removable light collar made of plastic, felt or woven material will limit flexion and extension and permit daily routine *activity. A cervical brace in which the* mandible and the occiput are engaged restricts rotation. Complete immobilization is secured by a Minerva cast in which the head, the neck and the chest are incorporated.

ESTROGENS. Cervical arthritis frequently occurs in women, *starting at the menopause.* Theoretically, use of estrogenic hormones is warranted. The patient feels better but it is doubtful whether the degenerative process is retarded.

PROCAINE INJECTIONS. Procaine may be injected into the soft tissues about the site of pain and tenderness. The beneficial effect probably results from temporary relief of muscle spasm and pain from the ligaments, permitting free motion which stretches the periarticular structures and reduces irritation. *Treatments are given twice weekly* until symptoms are relieved. Precautions are taken

to avoid inserting the needle too deeply. Barbiturates are given to prevent a procaine reaction.

Vaccines, artificial fever therapy, gold therapy, bee venom, *etc.*, *have no place in the treatment of degenerative joint disease.*

CONVALESCENT PERIOD. Activities which impose stresses and *strains on* the cervical spine should be avoided, *if necessary*, by changing the occupation. *Graded gentle exercises* strengthen the extensors and provide more stability. Small doses of salicylates, occasional application of heat, and periods of rest help to halt an attack in its incipient stage. A warm, dry climate reduces the symptoms but in itself does not prevent degeneration. Forceful manipulation of the spine should be avoided, as it tears the soft tissues and aggravates the symptoms.

Surgical Treatment. When localized degeneration follows fracture or dislocation and symptoms are persistent, a *spine fusion* between the affected vertebrae eliminates the disability.

TECHNIC Through a mid-line incision, the spinous processes, the articular facets and the laminae are exposed subperiosteally. The two vertebrae, having been identified by an x-ray marker, are curetted thoroughly, and the interspinous ligament is removed. Identification is aided by the prominent 2nd cervical and 7th cervical spinous processes. The last bifid spinous process is usually the 5th cervical. A rectangular corticocancellous iliac bone graft is laid in each gutter and is supplemented by chips. A wire suture tied about both spinous processes ensures stability. Postoperatively, head halter traction and sandbags control movement. Two weeks later, sutures are removed, and a Minerva cast is applied. Fusion is usually adequate at 4 or 5 months.

DECOMPRESSION LAMINECTOMY. This operation is performed for nerve root symptoms It is unnecesary to remove the offending spurs. Release of pressure relieves symptoms. Spine fusion may be added if necessary. (See section on Cervical Disk)

TORTICOLLIS
(Wryneck)

Torticollis is a rotational deformity of the cervical spine which secondarily causes turning, tilting and deformity of the head. The longer a wryneck is allowed to exist, the more resistant to treatment it becomes because of deformity developing in the individual bony components of the cervical spine, and contractures of the soft tissues. The following are the important causes of torticollis:

CONGENITAL TORTICOLLIS

The deformity is observed at birth and most often displays unilateral tightness of the soft tissues, especially the sternocleidomastoid muscle. Frequently, a fibrous tumor is found within the muscle. Rarely, the congenital deformity is a consequence of bony anomalies of the cervical spine, the Klippel-Feil syndrome. These conditions are discussed in the section on Congenital Deformities.

TRAUMA

A sprain, a dislocation or a fracture of the cervical spine often causes rotational deformity. A common condition is the so-called "subluxation." The patient gives a history of turning the head about to look backward and experiencing a severely painful snap in the neck. The cervical spine is locked in the rotatory position, muscles are in spasm, and a localized point of tenderness over the spine is found. Roentgenograms are noncontributory because the overlapping of rotated vertebrae precludes proper visualization. Traction under anesthesia suddenly reduces the deformity, with immediate relief of pain and restoration of motion.

Spontaneous subluxation occurring in children 6 to 12 years of age and often following upper respiratory infection is a common cause of "acquired torticollis."[8] About a week after the infection, the child complains of a "crick in the neck," and the head is held rigidly in the position of torticollis. However, spasm or contracture of the sternocleidomastoid is absent. Roentgenograms reveal the forward displacement of the atlas. Reduction is effected under general anesthesia, the head being immobilized in extension and neutral rotation in plaster.

The sudden onset of torticollis in a child or an adult, in spite of the absence of a history of trauma, should direct one's attention to the cervical spine. A paralyzing accident from unwarranted manipulation of an unsuspected

[8] Watson-Jones, R.: Spontaneous hyperemic dislocation of the atlas, Proc. Roy Soc. Med 25:58, 1932.

cervical displacement will be prevented thereby. Every case of torticollis deserves x-ray study before treatment is instituted.

Myositis or Fibromyositis

The common stiff neck which follows exposure to a cold draft causes painful tender cervical muscles, the nature of which is unknown. When involvement is unilateral, the patient holds the head toward that side to relax the muscles, the torticollis lasting only a day or two. Treatment consists of heat, preferably moist, recumbency, a soft collar and sedation.

Spasmodic Torticollis

The spontaneous appearance of painful, persistent or intermittent, muscle contractions producing the typical wryneck deformity is designated as spasmodic torticollis. The condition is resistant to the usual forms of therapy, including muscle sectioning, and these unfortunate individuals often develop severe psychiatric disturbances. The cause is unknown, but Foerster[9] explained the abnormally involuntary movements as resulting from an intermittent flow of thalamic impulses by way of the pallidum, following the occurrence of lesions of the corpus striatum.

Electromyographic studies of various muscles reveal *bilateral* involvement of many muscles, especially the sternocleidomastoid, the trapezius and the splenius. The spinal cord segments from the 1st cervical to the 1st thoracic supply all these muscles so that it is impossible to denervate these muscles completely by resection of their supposed nerve roots of supply. Treatment consists of intradural section of both spinal accessory nerves and the first 3 anterior cervical nerve roots. The procedure may be varied, depending upon the severity of involvement as indicated by

[9] Foerster, O : On indications and results of excision of posterior spinal roots in men, Surg, Gynec. & Obst 16.463, 1913.

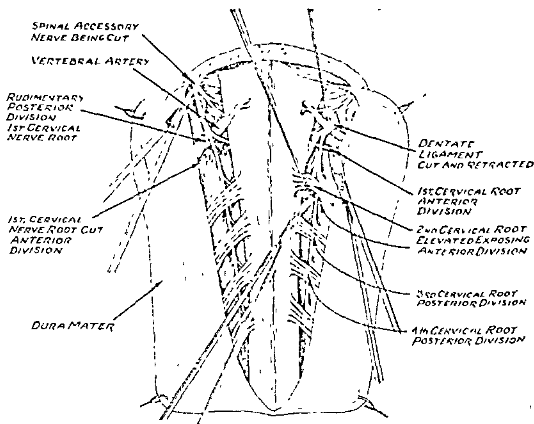


Fig. 283 Spasmodic torticollis. Exposure of the spinal cord and the medulla, showing intradural section of the spinal accessory nerves, anterior cervical nerve roots, and section of the dentate ligament to facilitate exposure of the first anterior cervical nerve root. (Wycis, H. T., and Moore, J. R.: The surgical treatment of spasmodic torticollis, J. Bone & Joint Surg. 36A:119)

electromyographic studies preoperatively. Minimal involvement on one side and severe involvement on the other may require only nerve root section on one side only. When pain is intense, bilateral, and many muscles are at fault, section of the 4th anterior cervical nerve root may be added without fear of compro-

Neuritis of the Spinal Accessory Nerve

The causes are similar to those causing neuritis elsewhere (see Multiple Neuritis). The spinal accessory nerve enters the neck at the lateral border of the upper third of the sternocleidomastoid at the level of the 2nd cervical vertebra. At this point the nerve is found to be tender, and injection of a local anesthetic temporarily relieves the painful muscle spasms. The condition is temporary and is treated by heat, rest, sedation, repeated local anesthesia and removal of the offending factor when found. Differentiation from true spasmodic torticollis is established by the involvement being unilateral and by relief of symptoms by injection of a single nerve.

Infections of the Cervical Spine

Tuberculosis is the chief offender. Destruction of several vertebrae, including the intervening disks, insidious onset and slow progression, a frequently associated retropharyngeal abscess and constitutional signs suggest the diagnosis.

Paralytic Torticollis

Unilateral cervical muscle involvement in poliomyelitis is rather unusual. When it does occur, the head is rotated toward the side of paralysis. When it can be established that the sternocleidomastoid is the chief offender, a muscle transfer procedure may be possible. Otherwise, fusion of the cervical spine, excluding the atlanto-axial joint, is indicated

Ocular Disturbances

This condition develops during childhood. No deformity is apparent at birth but develops as the child compensates for the abnormal

[10] Wycis, H T., and Moore, J. R.: The surgical treatment of spasmodic torticollis, J. Bone & Joint Surg. 36A 119, 1954



Fig. 284. Acute torticollis due to neuritis of the spinal accessory nerve. The severely painful spasms were unilateral, temporary and relieved by infiltration of the nerve with a local anesthetic. In contrast, spasmodic torticollis is bilateral, persistent or intermittent, and unaffected by injection of the nerve.

vision. Every case of torticollis which develops slowly in early childhood should be subjected to an examination of the eyes.

BONY ABNORMALITIES ABOUT THE CRANIOVERTEBRAL JUNCTION[11, 12]

Bony malformation about the foramen magnum constitutes an important cause of symptoms in the extremities, particularly the upper. Compression of the neuraxis, intramedullary cavitation (syringomyelia), and displacement of the cerebellum through the foramen magnum are possible associated soft-tissue pathology. The neurologic disturbances simulate and are often diagnosed as multiple sclerosis or syringomyelia. These malformations, in most cases, remain asymptomatic. However, a considerable number develop neurologic signs and symptoms in adolescence or adulthood, usually following an injury or unusual exercise. The

[11] McRae, D. L Bony abnormalities in the region of the foramen magnum. Correlation of anatomic and neurologic findings, Acta radiol. 40:335, 1953.

[12] Spillane, J. D, Pallis, C., and Jones, A. M: Developmental abnormalities in the region of the foramen magnum, Brain 80.11, 1957.

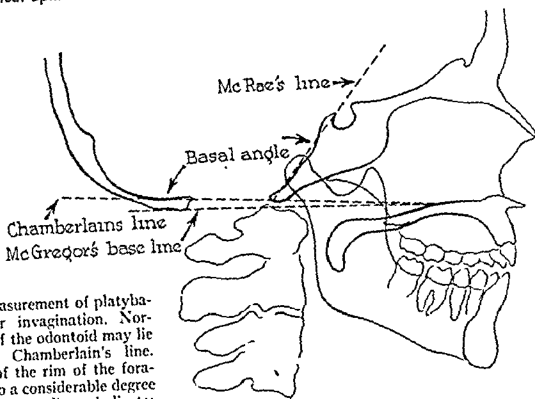


FIG. 285. Measurement of platybasia and basilar invagination. Normally, the tip of the odontoid may lie slightly above Chamberlain's line. Displacement of the rim of the foramen magnum to a considerable degree above McGregor's line indicates basilar impression. McRae's line lies along the plane of the clivus. It normally subtends an angle with Chamberlain's or McGregor's base line not exceeding 145°. A more obtuse angle indicates platybasia. (McRae, D. L., and Barnum, A. S.: Am. J. Roentgenol. 70:23; McGregor, M.: Brit. J. Radiol. 21:171)

disability is characteristically transient and recurring, predominantly affecting the upper extremities, and occasionally assuming serious proportions, e.g., a quadriplegia. This combined orthopaedic and neurologic problem demands careful study and recognition of bony abnormalities about the craniovertebral junction. Decompression of the foramen magnum and the 1st cervical vertebra, often combined with occipitocervical fusion, will greatly reduce and occasionally eliminate completely the neurologic disability. The following types of malformations may exist individually or in combination.

Basilar Invagination or Impression

Basilar impression is an upward bulging of the rim of the foramen magnum. It represents (1) a congenital maldevelopment or (2) a deformity secondary to bony softening, e.g., Paget's disease. When it occurs as an isolated lesion without other neighboring malformation, it is usually asymptomatic. However, signs of syringomyelia or syringobulbia are often associated. Prolapse of the cerebellum through the foramen (cerebellar ectopia) commonly is associated with basilar impression.

Roentgenologic Signs of Basilar Impression:

1. Upward curvature of the lips of the foramen

2. The foraminal rims, and often the odontoid tip, lie above the base lines:

A. *Chamberlain's line:* from hard palate to inner aspect of squamous occipital cortex (lateral view)

B. *McGregor's line:* from hard palate to external aspect of squamous occipital cortex (lateral view)

C. *Fischgold's line:* line passes through tips of mastoid processes (anterior transoral view)

Symptoms and signs are likely when the odontoid tip lies well above the base line. However, a high odontoid is not always an indication of an elevated foraminal rim; it may represent (1) an abnormally long odontoid, or (2) occipitalization of the atlas (q.v.).

One must remember that the basal occiput slopes gently upward and backward so that the posterior rim of the foramen magnum always lies at a slightly higher level than the anterior.

The literature often implies that basilar



FIG. 286. Occipitalization of the atlas. Arrows point to the assimilated anterior and posterior arches. (McRae, D. L.: Bony abnormalities in the region of the foramen magnum, Acta radiol. 40:335)

impression is synonymous with platybasia. However, platybasia, defined as flattening of the base of the skull, is an entirely different entity. It is without clinical significance. It can be determined by ascertaining an angle subtended by the plane of the clivus and Chamberlain's or McGregor's line. When this exceeds 140° (approaching 180°), platybasia is said to exist.



FIG. 287. Occipitalization of the atlas—midsagittal laminogram.

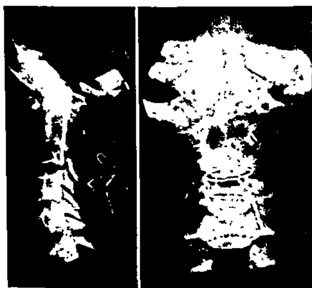


FIG. 288. Occipitalization of the atlas—postmortem specimen. (Same case as Figs. 286 and 287.)



Fig. 289. Separate odontoid process with marked gliding of the atlas on the axis evident in flexion of the neck. Spastic weakness of all extremities. (McRae, D. L.: Bony abnormalities in the region of the foramen magnum, Acta radiol. 40:335)



Fig. 290. Chronic atlanto-axial dislocation.

Occipitalization of the Atlas
(Assimilation of the Atlas to the Base of the Skull; Atlanto-occipital Fusion)

Bony union of varying degree may exist between skull and atlas. The atlas may be partially (an arch, a lateral mass) or completely assimilated into the skull. Usually, it is associated with other bony abnormalities about the foramen magnum, particularly basilar impression and chronic atlanto-axial dislocation. In compensation for absolute atlanto-occipital immobility, excessive mobility is demonstrable between atlas and axis. A Klippel-Feil type of syndrome is often an associated condition.

Any degree of atlanto-occipital fusion, partial or complete, will prevent all motion at this junction. Normally, on a lateral x-ray film with the neck in full flexion, the posterior arch separates from the overlying squamous occiput. Failure of separation defines occipitalization of the atlas.

Neurologic signs and symptoms are likely if:

1. The odontoid is long, high, or angulated posteriorly.

2 A-P diameter of spinal canal behind the odontoid is 19 mm. or less, especially when a gap of more than 3 mm. separates the odontoid from the anterior arch.

3. Odontoid movement is more than 3 mm. on flexion and extension of head.

4. The foramen magnum diameter may be reduced by a bony prominence posteriorly, representing an assimilated posterior arch of the atlas.

Most cases with neurologic disability have a chronic atlanto-axial dislocation.

Chronic Atlanto-Axial Dislocation

This is a congenital abnormality which may be due to:

1. Congenital absence of the odontoid.
2. Failure of the odontoid to fuse with the body of the axis. In this case, the odontoid is often fused to the anterior arch of the atlas.
3. Absence or abnormal laxity of the transverse ligament.

Most frequently, one observes in roentgenograms a normal odontoid, so that a deficiency of the transverse ligament is presumed. Little or no atlanto-axial gliding takes place with flexion and extension movements of the neck. Occipitalization of the atlas is often associated. A lateral roentgenogram with the neck in full flexion is necessary to define this condition.

Chronic atlanto-axial dislocation is particu- [illegible]

spontaneous atlanto-axial dislocation of childhood. The latter usually follows an upper respiratory infection, seldom produces neurologic symptoms, is easily reduced and practically never recurs.[13]

Separate Odontoid Process of Axis

This is a congenital anomaly. The dens is unusually small in length and breadth and is often fused to the anterior arch of the atlas or the anterior rim of the foramen magnum. It permits forward gliding of the atlas on the axis during flexion and extension of the head and the neck. In roentgenograms the odontoid is observed to move with the anterior arch of the atlas.

In the adult, occasionally it is difficult to ascertain whether a separate odontoid is congenital or ununited following fracture. Nonetheless, the effect is the same.

Fusion of Cervical Vertebrae

This may vary from fusion of 2 vertebrae to extensive fusions with lessening in number of cervical vertebrae. Characteristic features of the Klippel-Feil syndrome (q.v.) may be associated and include short neck and low hairline. The Klippel-Feil syndrome in itself produces no neurologic disability. However,

[13] Watson-Jones, R.: Spontaneous hyperemic dislocation of the atlas, Proc. Roy. Soc. Med. 25.58, 1932.



Fig. 291. Atlanto-axial anterior dislocation demonstrating the degree of cord compression in: (*left*) displacement with an intact odontoid and (*right*) displacement with a fractured odontoid. The former, whether spontaneous, traumatic or congenital, is extremely serious.



FIG. 289. Separate odontoid process with marked gliding of the atlas on the axis evident in flexion of the neck. Spastic weakness of all extremities. (McRae, D. L.: Bony abnormalities in the region of the foramen magnum, Acta radiol. 40:335)



FIG. 290. Chronic atlanto-axial dislocation.

OCCIPITALIZATION OF THE ATLAS

(Assimilation of the Atlas to the Base of the Skull; *Atlanto-occipital Fusion*)

Bony union of varying degree may exist between skull and atlas. The atlas may be partially (an arch, a lateral mass) or completely assimilated into the skull. Usually, it is associated with other bony abnormalities about the foramen magnum, particularly basilar impression and chronic atlanto-axial dislocation. In compensation for absolute *atlanto-occipital* immobility, excessive mobility is demonstrable between atlas and axis. A Klippel-Feil type of syndrome is often an associated condition.

Any degree of atlanto-occipital fusion, partial or complete, will prevent all motion at this junction. Normally, on a lateral x-ray film with the neck in full flexion, the posterior arch separates from the overlying squamous occiput. Failure of separation defines occipitalization of the atlas.

Neurologic signs and symptoms are likely if:

1. The odontoid is long, high, or angulated posteriorly.

2. A-P diameter of spinal canal behind the odontoid is 19 mm. or less, especially when a gap of more than 3 mm. separates the odontoid from the anterior arch.

20 6/10/90

Cervicobrachial Region

PATHOLOGY OF CERVICOBRACHIAL COMPRESSION[1]

Symptoms which are predominantly in the hand are caused by a hang-up of the lowest trunk of the brachial plexus on an abnormal structure. More widespread symptoms, such as pain in the arm, point to compression of the plexus between the clavicle and an abnormal structure, most commonly a cervical rib. This occurs during retraction and abduction of the shoulder, which causes the clavicle to pivot on its medial end, elevating and rotating backward toward the base of the neck.

A cervical rib, or a fibrous band representing the rib, attaches to the anterior end of the 1st thoracic rib and narrows the interval through which pass the brachial plexus and the axillary artery. With advancing age, the shoulder droops, the scalenus anterior elongates, the angle between the cervical rib and the scalenus anterior becomes more acute, and

[1] Telford, E. D., and Mottershead, S.: Pressure at the cervicobrachial junction, J. Bone & Joint Surg. 30B.249, 1948.

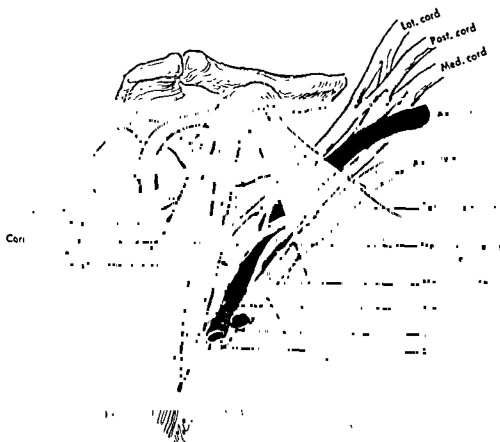


FIG. 292. Relationships of neurovascular structures pectoralis minor and the second part of the axillary artery. to this, the radial and the axillary nerves lie behind the a structures lie on a floor formed by the subscapularis pro teres major and the latissimus dorsi distally.

oft-associated malformations about the foramen magnum are particularly susceptible to trauma in view of immobility of the rest of the cervical spine.

Clinical Picture

Malformations in most instances are asymptomatic and are discovered by chance in roentgenograms; or symptoms and findings develop after trauma or following a period of vigorous unaccustomed exercise. The neurologic disability is extremely variable in extent and degree, usually is characteristically pronounced in the upper extremity, and is transient.

Symptoms consist mainly of weakness, ataxia, numbness and pain in the arms and the legs. Neck pain is frequent, located in the upper neck and the occiput, not related to head movements, and is relieved by lying down. It is not the sharp, shooting type of pain which is typical of greater occipital neuralgia. Symptoms of increased intracranial tension are seldom experienced, although headache and dizziness are common.

Neurologic signs include mainly the following:

1. Corticospinal tract deficit in arms and legs (hyperreflexia, Babinski, etc.)
2. Ataxia in arms and legs
3. Nystagmus, often associated with subjective sensation of oscillating objects (oscillopsia). Therefore, patients have a dislike for descending stairs. Due to herniation of cerebellar tonsils.
4. Bulbar signs
5. Posterior column deficit: proprioceptive loss, 2-point astereognosis. When confined to upper limbs, it constitutes a characteristic feature of cord compression at the foramen magnum.
6. Syringomyelic sensory disorders
7. Signs of increased intracranial pressure are rare.

Clinical appearance of the patient is usually normal. The following suggest the presence of an anomaly, especially occipitalization:

1. Unusual posture of head
2. Abnormal configuration of neck
3. Restricted mobility of neck
4. Altered position of hairline
5. Short neck.

When the Klippel-Feil syndrome is associated with symptomatic malformations, "mirror movements" are often a characteristic. These consist of one extremity duplicating the movements of the other, e.g., extension of the index finger of one hand is accompanied immediately by extension of the opposite index finger. Dissociated movements such as are necessary for climbing stairs are impossible.

Laboratory Diagnosis

The procedures utilized are:

1. *Myelography* (lumbar route), demonstrating a block at the foramen magnum, a cervical syrinx, and ectopia of cerebellar tonsils.
2. *Encephalography*, demonstrating ventricular enlargement; failure to fill often associated with cerebellar ectopia.
3. *Vertebral Angiography*. The posterior inferior cerebellar artery enters the upper part of the spinal canal and retraces its course to the cranial cavity. This is the "loop sign" of cerebellar ectopia.

Regular roentgenograms may be sufficient, but planograms (tomograms) in the lateral midsagittal and A-P midfrontal plane will display the bony abnormality more clearly.

Treatment[14, 15]

Severe and progressive neurologic disability requires surgical intervention. Cases of "multiple sclerosis" and "syringomyelia" should be subjected to intensive study of the region of the foramen magnum. Decompression of the foramen magnum and the posterior arch of the atlas often produces a dramatic remission of symptoms. Particularly when atlanto-axial dislocation is present, a cervico-occipital fusion is added. The loss of motion when the occiput is fused to the 2nd cervical vertebra is no more than 50 per cent. Fusions extended to the 3rd cervical vertebra result in loss of about 90 per cent of motion. Postoperatively, a light brace is worn and followed, after 2 weeks, by a plaster cast for about 16 weeks, or until roentgenograms show a satisfactory fusion.

[14] Lipscomb, P. R.: Cervico-occipital fusion for congenital and post-traumatic anomalies of the atlas and axis. J. Bone & Joint Surg. 39A:1289, 1957.

[15] Cone, W., and Turner, W. G.: The treatment of fracture dislocation of cervical vertebrae by skeletal traction and fusion. J. Bone & Joint Surg. 19:584, 1937.

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Cervicobrachial Region

PATHOLOGY OF CERVICOBRACHIAL COMPRESSION[1]

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[1] Telford, E. D., and Mottershead, S.: Pressure at the cervicobrachial junction, J. Bone & Joint Surg. 30B 249, 1948

This occurs during retraction and abduction of the shoulder, which causes the clavicle to pivot on its medial end, elevating and rotating backward toward the base of the neck.

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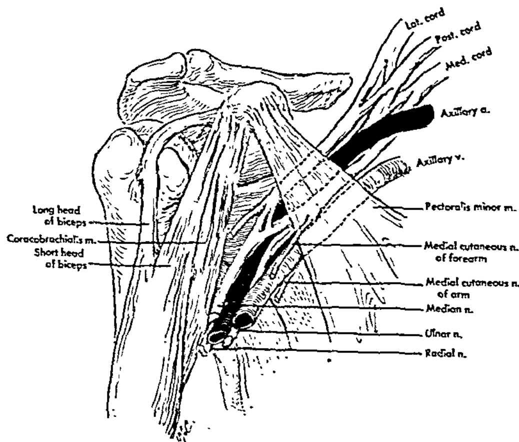


FIG. 292. Relationships of neurovascular structures at the level of the pectoralis minor and the second part of the axillary artery. Immediately distal to this, the radial and the axillary nerves lie behind the axillary artery. All structures lie on a floor formed by the subscapularis proximally and the teres major and the latissimus dorsi distally.

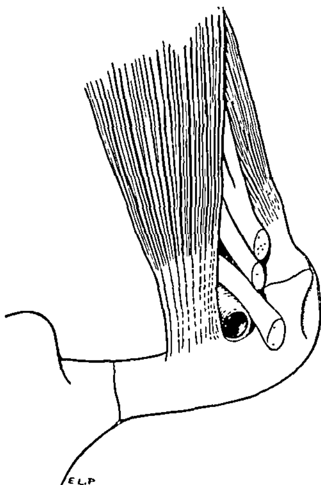


Fig 293 Compression of the subclavian artery and inferior trunk of the plexus between scalenus anterior and the forward end of a cervical rib. (Telford, E. D , and Mottershead, S.: Pressure at the cervico-brachial junction, J. Bone & Joint Surg 30B:249-265)

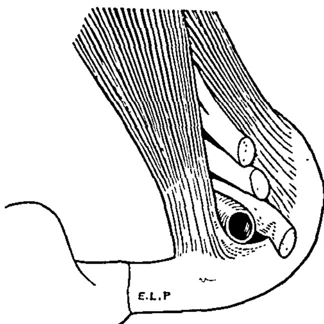


Fig. 294. The rare falciform type of insertion of scalenus anterior. (Telford, E. D., and Mottershead, S.: Pressure at the cervico-brachial junction, J. Bone & Joint Surg. 30B:249-265)

the neurovascular bundle becomes compressed The lowermost trunk of the plexus is hung up over the rib, causing a notch in the trunk and a groove in the rib. This situation is accentuated by a postfixed plexus The lower fibers must cross the cervical rib at an acute angle to reach the lower trunk of the plexus In addition, the 1st thoracic nerve, which runs in the first intercostal space, must ascend in front of the neck of the 1st rib and pass over the upper border of the rib to join the lower trunk.[2] Fibers in this trunk originate chiefly from the 8th cervical and the 1st thoracic nerve roots; therefore, pain and paresthesias are limited to the ulnar side of the hand. The trunk also relays sympathetic fibers to perivascular tissues. Vasospasm causes ischemic symptoms such as pallor, cyanosis, muscle cramps, particularly with activity, and even gangrene Vasoconstriction may lead to extensive thrombosis. When the axillary artery is constricted in the narrow rib-muscle interval, an aneurysm forms immediately distal to the point of constriction.

The most common cause of compression is a cervical rib. When a small, underdeveloped rib is present, it continues forward as a fibrous band to attach to the 1st rib adjacent to the lateral edge of the tendon of the scalenus anterior. At this point a large exostosis may develop and contribute to the compression. The lateral portion of the tendon may be extended laterally as a sharp, falciform edge which cuts into the artery and the lowermost trunk

In the absence of a cervical rib or band, compression may occur between the tendinous insertions of the scalenus anterior and the medius This occurs particularly when the tendons are broad, falciform and overlapped. This is the type relieved by section of the scalenus anterior.

[2] Williams, A F.: Role of the first rib in scalenus anticus syndrome, J. Bone & Joint Surg 34B 200, 1952

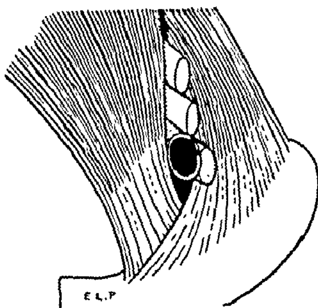


Fig. 295 The V type in which the insertions of scalenus anterior and scalenus medius overlap. (Telford, E. D., and Mottershead, S.: Pressure at the cervicobrachial junction, J. Bone & Joint Surg. 30B 249-265)

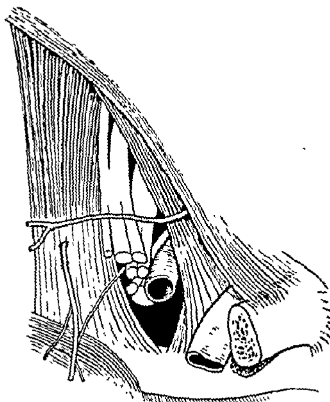


Fig. 296. The normal pattern of insertion of the scalenus anterior and scalenus medius. (Telford, E. D., and Mottershead, S.: Pressure at the cervicobrachial junction, J Bone & Joint Surg 30B:249-265)

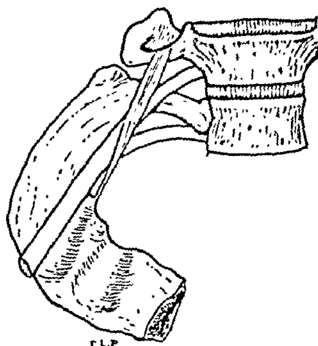


Fig. 297. Scalenus minimus (muscle of Albinus) compressing the first thoracic nerve root. (Telford, E. D., and Mottershead, S.: Pressure at the cervicobrachial junction, J. Bone & Joint Surg. 30B:249-265)

About 35 per cent of cases have a muscle band (muscle of Albinus, scalenus minimus) which arises from the transverse process of the 7th cervical vertebra and ends in a slender, rounded tendon attached to a small spur on the inner margin of the 1st rib at a point between the two scaleni. The muscle belly crosses and compresses the 1st thoracic nerve at the neck of the 1st rib. This muscle should be sought and sectioned *after* scalenotomy has been done.

Scalenus anticus spasm is not a cause of cervicobrachial compression. Contraction of the muscle would, in effect, elevate the 1st rib and widen the neurovascular interval. Conversely, weakening of the muscle allows the rib to descend, and compression results from passiveness of the muscle.

Costoclavicular compression can occur only when the costoclavicular interval is narrowed by a large cervical rib or an abnormal first thoracic rib. The latter most commonly is associated with thoracic scoliosis. Clavicular pressure can occur only during retraction of the shoulder.

When symptoms of pain, weakness and

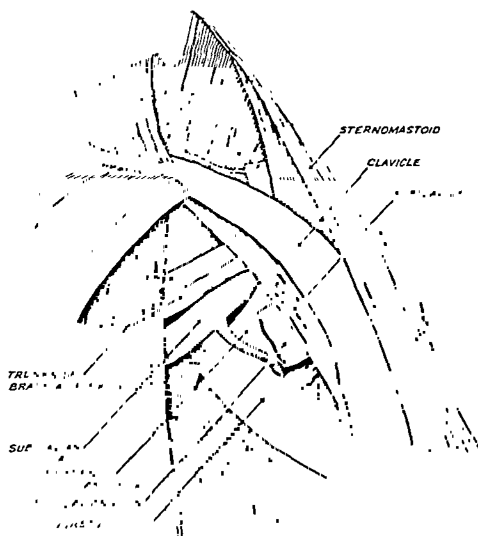


FIG. 298. Illustrating how rotation of the clavicle in its long axis causes the anterior convexity of its medial two-thirds to form an arch over the neurovascular bundle (arm abducted to 90°). (Telford, E. D., and Mottershead, S.: Pressure at the cervico-brachial junction, J. Bone & Joint Surg. 30B:249-265)

numbness occur with depression of the shoulder, e.g., while supporting a heavy weight, diminution or obliteration of the radial pulse attests to vascular compression as the cause. The axillary artery distal to the clavicle and at the level of the 2nd rib turns laterally and passes between two nerve trunks which form the median nerve. Beyond this point it turns again distally to proceed down the arm. Traction downward on the arm causes the artery to be constricted between the median nerve trunks. The tortuous course of the artery provides extra length necessary for abduction at the shoulder. Occasionally, the artery may be stretched and irritated beneath the coracoid by hyperabduction movements giving rise to symptoms of ischemia. If, while the arm is abducted, an effort is made to adduct the arm against resistance, the artery may be compressed between the pectoralis minor and the subscapularis.

CERVICAL RIB

The cervical rib is a supernumerary rib which arises usually from the 7th, rarely from the 6th or the 5th, cervical vertebrae. It is frequently bilateral.

Developmental Anatomy

In the embryo, the nerves are much larger in proportion to the ribs than they are in the fully developed animal. When the nerves are unusually large, as they are in the cervical region, they interfere with the development of costal processes. The brachial plexus has two distinct types of arrangement.

The *prefixed plexus* has a well-developed 4th cervical root and a small 1st thoracic root. Formation of a costal process encounters little resistance from the small 1st thoracic nerve root. As a result, there develops a rib extending from the transverse process of the 7th cervical vertebra varying in size from a small

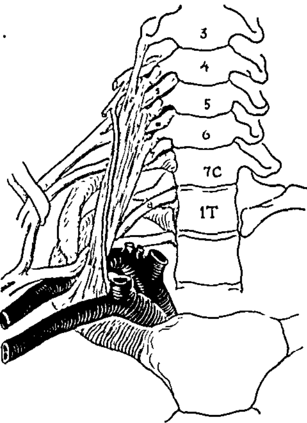


Fig. 299. Cervical rib. The anterior portion or its counterpart, a fibrous band, compresses the lower cord of the brachial plexus and the subclavian artery against the scalenus anticus. Often the tendinous insertion of the scalenus anticus extends proximally and narrows the rib-scalenus interval. Or the cervical rib extends far forward, and the neurovascular structures are "hung up." Note the phrenic nerve coursing along the anterior surface of the muscle.



Fig. 300. Cervical rib. The small bony process often overlies the proximal portion of the first thoracic rib and therefore is easily overlooked. The interval between the distal end of the cervical rib and the anterior end of the thoracic rib (scalene tubercle) is bridged by a dense fibrous band.

rudimentary rib to a complete rib extending forward to articulate with the sternum or the costal cartilage of the 1st thoracic rib. Where the rib is underdeveloped, a fibrous band may extend from its outer extremity to the 1st thoracic rib, usually attaching near the scalene tubercle Intercostal muscles, nerve and vessels are usually associated with a completely formed cervical rib.

A *postfixed plexus* receives few fibers from the 4th cervical root but has a large, well-developed 1st thoracic nerve root. Formation of a costal process is less likely.

Pathologic Anatomy

Regardless of whether the rib is fully developed or represented by a fibrous band, the brachial plexus and the subclavian vein must pass over a higher barrier before passing downward to the arm. The neurovascular structures are hung up. In addition, the cervical rib or band inserts anteriorly at or near the scalene tubercle, thereby narrowing the interval through which nerves and artery pass. At the point of insertion, the tubercle on the first thoracic rib may become enlarged and add to the compression and friction. Plexus and artery are further embarrassed when they are pulled distally by downward traction on the arm, such as when carrying a heavy weight. Normally, with advancing age, the shoulder girdle droops dow[illegible] eases tension [illegible] Pronounced [illegible] in women of middle age, in the course of unusual lifting occupations, and following an acute illness when muscle weakness develops. This explains the frequency of symptoms in these situations.

The lowermost cord of the plexus is subject

to the greatest degree of friction and compression. It contains sensory fibers to the ulnar distribution, motor fibers to the intrinsic muscles of the hand, and vasomotor fibers to vessels beyond the level of the tendon of the pectoralis major.

A cervical rib narrows the thoracic outlet. As a result, both plexus and artery may be compressed between the clavicle and the rib during elevation and retraction of the arm.

Clinical Picture

Symptoms can occur at any age but often are initiated under conditions effecting descent of the shoulder girdle. Symptoms and findings are characteristically ulnar in distribution, pointing to the lower trunk of the plexus. Complaints referable to the median nerve distribution usually implicate a ruptured cervical disk.

Symptoms

Pain and paresthesias occur in the ulnar aspect of the hand and the little and the ring fingers. Less commonly, it may be felt in the whole hand. The pain may be dull and aching or sharp and lancinating. A sensation of tingling of the forearm and the hand, which the patient describes as "falling asleep," is ascribed to circulatory deficiency and is associated with diminution of the radial pulse. Weakness of the hand, clumsiness in use of the fingers, and dropping of objects is complained of. Symptoms are accentuated by downward displacement of the shoulder girdle, e.g., when carrying a heavy object or following the fatigue brought on by excessive activity. *Adson's sign*[3] intensifies the symptoms by increasing tension on the scalenus anterior and narrowing the rib-muscle interval. The patient rotates the chin toward the affected side, elevates the chin and hyperextends the neck, and then takes a deep breath. This increases pain and paresthesia and often obliterates the radial pulse.

Findings

It may be possible to feel the bony prominence of the cervical rib at the base of the neck. *Failure to palpate the rib does not rule out this syndrome*, as a fibrous band may be the offender. The irritated plexus is tender to deep pressure lateral to and behind the sternocleidomastoid. The muscles supplied by the lower trunk *may be atrophied*. *Most commonly*, these are the interossei and the lumbricals. Flexion at the metacarpophalangeal joints, extension at the interphalangeal joints, abduction and adduction of the fingers, and adduction of the thumb are weak. Less commonly, the thenar muscles, especially the abductor pollicis brevis and the opponens pollicis, are involved. Sensation is diminished over the ulnar border of the forearm and the volar aspect of the little finger and the ulnar half of the ring finger. Diminished circulation is evidenced by coldness, pallor, cyanosis, and reduced volume of the radial pulse. The subclavian artery above the clavicle and the axillary artery below the clavicle are felt to be pulsating, indicating that vasospasm has occurred distally in the brachial artery. Trophic changes include a thin glossy skin, ulcerations, and ridging and brittleness of the nails. Localized areas of gangrene indicate that thrombosis of a main artery has occurred, usually in the lower region of the arm.

Roentgenologic Findings. A cervical rib is found extending outward from the 7th cervical transverse process. It may be small and rudimentary or large and fully developed. The *condition is often bilateral*.

Differential Diagnosis

The presence of a cervical rib does not necessarily implicate it. Many cervical ribs do not produce symptoms.

A herniated cervical disk is most frequently confused with cervicobrachial compression. In this condition, the 5th cervical disk is usually involved, causing symptoms and findings in the median distribution of the hand There is tenderness over the cervical interspace; hyperextending the neck increases the pain; and downward pressure of the head on the cervical spine accentuates the symptoms. The radial pulse is not affected. At a later stage, the disk space becomes narrowed, and localized degenerative changes supervene.

Costoclavicular compression *is identified* by intensification of symptoms when the arm is adducted and retracted. A cervical rib or a deformed 1st thoracic rib is present.

[3] Adson, A. W., and Coffey, J. R Cervical rib, Ann Surg. 85:839, 1927.

The hyperabduction syndrome is identified by diffuse nerve symptoms throughout the extremity and obliteration of the pulse produced by the hyperabducted position. Symptoms are accentuated when an attempt is made to adduct the arm from the abducted position against resistance.

Other conditions to be ruled out are ulnar nerve lesions, syringomyelia, and tumors within the spinal canal.

Treatment

Theoretically, elevation of the shoulder girdle releases tension on the brachial plexus and the axillary artery. When symptoms are minimal, conservative treatment may be tried. This consists essentially of exercises designed to increase the tone of the trapezius and the levator scapulae. The arm is placed at rest in the elevated position. Activities requiring lifting are avoided. Often symptoms are severe, and surgical removal of the rib is necessary.

SCALENUS ANTICUS SYNDROME

Cervicobrachial compression can occur in the interval between the scalenus anterior and the scalenus medius in the absence of a cervical rib. Each muscle may have a large tendon and a widespread insertion even to the point of overlapping so that the interval is narrowed and the plexus and the artery are hung up. The edge of the tendon may extend forward from the scalenus medius and backward from the scalenus anterior as a sharply edged falciform structure which cuts into the artery and the lower trunk of the plexus. The scalene tubercle often forms a large bony prominence which elevates the neurovascular structures. Clinically, the picture is identical with that produced by a cervical rib. The condition is often ascribed in the literature to spasm and hypertrophy of the scalenus anterior and is identified by relief of symptoms by local anesthesia of the muscle, relieving symptoms. However, overaction of the muscle would tend to elevate the rib and widen the interval. Paralyzing the muscle by local anesthesia would drop the rib and narrow the interval. The anesthetic very probably effuses into the subjacent plexus and thereby relieves symptoms. More probable is the abnormal insertions of the scaleni which traumatize the artery and the nerves. Treatment consists of sectioning the scaleni and, if necessary, excising the bony prominence.

Technic for Scalenotomy and Removal of Cervical Rib

A transverse incision is made just above the medial third of the clavicle. It is deepened through the platysma, severing the supraclavicular nerves. The external jugular vein is divided or retracted. The clavicular attachment of the sternocleidomastoid is divided, exposing fatty tissue covering the scalenus anterior. The subclavian vein is located inferiorly and must be protected. The transverse cervical artery is ligated and divided. By blunt dissection through the fat pad, the tendon of the scalenus anterior is exposed where it inserts on the 1st rib. The phrenic nerve runs along the anterior surface of the muscle and must be exposed as a precaution against injury. Immediately beyond the lateral border of the tendon, the pulsation of the subclavian artery is palpated. A blunt dissector is inserted between the artery and the posterior surface of the tendon. If the omohyoid, which crosses the muscle obliquely, interferes with exposure, it may be severed and resutured later. The scalenus tendon is severed, and it retracts, exposing artery and plexus to view. The upper two trunks of the plexus are retracted upward, and the lower trunk downward, exposing the anterior attachment of the cervical rib or the ligamentous band to the 1st rib. A band need only be sectioned. The rib must be followed backward carefully by blunt dissection and is completely removed. At this point, one should determine the presence of the scalenus minimus (muscle of Albinus) which inserts on the medial border of the 1st rib near the scalenus attachment. This must be severed. Through this same approach, a deformed 1st thoracic rib can be removed if costoclavicular compression is observed. The omohyoid, the sternocleidomastoid and the platysma are resutured, and the skin is closed.

BRACHIAL PLEXUS INJURIES

Anatomy[4]

The brachial plexus lies in the lower part of the posterior triangle of the neck, behind

[4] Cunningham's Manual of Practical Anatomy, New York, Oxford Univ Press, 1948.

the clavicle, and in the upper part of the axilla. The anterior primary rami unite just before the plexus emerges from behind the lower portion of the scalenus anterior. The lower trunk of the plexus is situated in the angle between the 1st rib and the scalenus anterior, which attaches to a tubercle on the rib anteriorly. In this acute interval, the scalenus medius lies above and behind, and the subclavian artery below and in front of the lower trunk. It is easy to see how the artery and nerve structures can be compressed between two actively contracting structures, or between the scalenus anterior and the bony rib.

The brachial plexus is formed by the anterior rami of the 5th, the 6th, the 7th and the 8th cervical, and the 1st thoracic nerves. When the plexus is well developed cranially, it receives an important contribution from the 4th cervical and is described as a prefixed plexus. A postfixed plexus is better developed caudally and is reinforced from the 2nd thoracic. The important 1st thoracic ganglion connects with the 1st thoracic ramus through which sympathetic fibers are carried from the spinal cord to half of the face. Therefore, injury to this area produces Horner's syndrome. The upper trunk is formed by union of the 5th and the 6th cervical rami, the lower trunk by the 8th cervical and the 1st thoracic, and the middle trunk by the 7th cervical alone. Each trunk then divides into anterior and posterior divisions which unite to form lateral, posterior and medial cords, describing their relationship to the second portion of the axillary artery behind the pectoralis minor muscle. The medial cord gives off the ulnar nerve and contributes the inner half of the medial nerve. The lateral cord gives off the musculocutaneous (supplies brachialis and biceps) nerve and contributes the outer half of the median nerve. The posterior cord gives off the axillary nerve, then continues distally as the radial nerve.

In general, the upper or the lateral portion of the plexus supplies muscles of the scapula, the shoulder and the flexor aspect of the arm The medial portion of the plexus controls the intrinsic muscles of the hand and the cer-

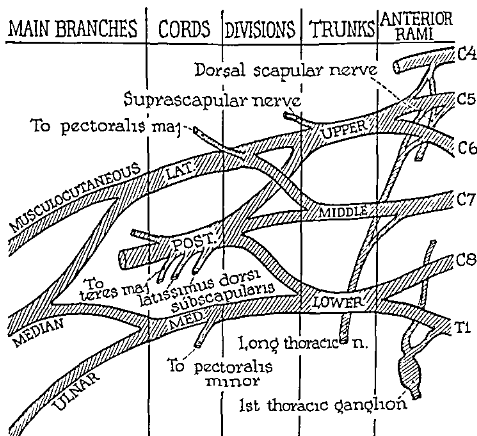


FIG. 301. The brachial plexus.

vical sympathetic. The posterior cord controls the deltoid and the extensors of the arm and the forearm.

The following are also of importance in localizing lesions:

Long thoracic (to serratus anterior) arises from the 5th, the 6th and the 7th cervical rami immediately after their exit from the intervertebral foraminae.

Dorsal scapular (to rhomboid and levator scapulae) arises from the 5th cervical ramus just lateral to the long thoracic.

Suprascapular (to supraspinatus and infraspinatus) arises from the lateral side of the upper trunk, well above the clavicle, and is the first important branch one sees when the plexus is explored above the clavicle.

Upper subscapular, lower subscapular (to subscapularis) **and thoracodorsal** (to latissimus dorsi) arise from the posterior cord.

Pathogenesis

The brachial plexus is injured by:

1. **Traction or Overstretching.** The head is flexed laterally toward the opposite side, and the shoulder is depressed. This is the mechanism in birth trauma and breech delivery. In shoulder dislocations where the arm is pulled violently out of the socket, traction on the brachial plexus is severe. Rupture of nerve fibers can occur at any point and can vary from a mild overstretching of the individual fibrils within their sheaths to tearing apart within the nerve trunks, cords, or peripheral nerves. When nerve roots are avulsed from the cord and through the intervertebral foramina, the meninges are torn away. The membranes then heal with formation of an empty meningocele which extends through the foramen. Rupture of nerve fibers is most frequent in that portion of the plexus which is subjected to the greatest stretch, i e., C5 and C6. Actual avulsion is rare.

2. **Compression.** Fracture of the clavicle and its callus formation, infection and subsequent scar contracture, tumors, hemorrhage and consequent cicatrix, and a direct blow to the side of the neck are causes.

3 **Penetrating wounds,** including stab or gunshot. The severance is usually in a well-defined place, and interruption of function is immediate. The concussion of a missile exerts its force over a widespread area and effects a temporary physiologic cessation of function far beyond the actual point of destruction.

Diagnostic Clinicopathologic Correlation

Before any treatment can be instituted, determine the extent of paralysis and then place the lesion by knowing the innervation of the involved muscles. First, it is wise to wait a short period to allow for spontaneous recovery of the physiologic lesion, particularly after a gunshot injury. One should not try to catalogue an injury into only an upper (Erb) or lower (Klumpke) type. The following are points which will enable a differentiation to be made:

1. Probable Site Determined by Mechanism of Injury

A. Stretching Rupture. As in birth injury, it occurs most frequently in that portion of the plexus which is subjected to the greatest stretch, namely, the 5th and the 6th roots, the upper trunk and the lateral cord. The lower brachial plexus, originating from C7, C8 and T1, is injured less often, and the degree of injury is generally less so that spontaneous recovery is more probable.

B. Stab or Bullet Wounds. Usually, the most exposed portions are injured, i.e., C5, C6, C7 and the upper trunk of the plexus.

C. Axillary or Infraclavicular Wounds. The individual nerves and the large blood vessels are more likely to be injured.

D. High-Velocity Missiles. By their concussion effects they produce paralysis over a variable wide area which subsides after a short interval.

2. Anatomic Grouping of Paralysis

A. Injury to spinal nerves at the foramina or trunks causes a segmental distribution of motor and sensory loss. The paralyzed muscles in a group correspond to the nerve root innervation:

C5, C6 and upper trunk—shoulder girdle and upper part of the extremity

C7, C8, T1 and middle and lower trunks —forearm and hand

T1 root or spinal nerve—Horner's syndrome

More specifically, the nerve roots innervate the following muscles:

C5—rhomboids, deltoid, supraspinatus,

infraspinatus, biceps, brachialis, clavicular head of pectoralis major

C6—sternal head of pectoralis major and triceps

C7—extensors of wrist and fingers

C8—flexors or wrist and fingers

T1—intrinsic muscles of hand and cervical sympathetic

The following nerves originate from the anterior primary rami shortly after exit of the latter from the foramina. Paralysis of their muscles of innervation places the lesion close to the spinal cord:

Long thoracic, to the serratus anterior, originates from C5, 6, 7; paralysis results in winging of the scapula and inability to push forward, and failure of fixation of the scapula during abduction at the glenohumeral joint.

Dorsal scapular, to the rhomboids and the levator scapulae, originates from C5 and C6 These muscles are important in fixing and rotating the scapula for scapulohumeral motion but are difficult to test individually.

Sympathetic fibers to the first thoracic ganglion originate from T1 ramus and are the main source of sympathetic supply to half of the face. Injury at this point produces a *Horner's syndrome.* This is characterized by unilateral findings of drooping upper eyelid, narrow palpebral fissure, enophthalmos, contraction of pupil, loss of ciliospinal reflex, and absence of sweating in the entire upper extremity—upper chest, neck and face

B. Injury to Cords. A more peripheral nerve pattern is produced.

Lateral Cord. The musculocutaneous (biceps) and upper half of the median nerve (flexor carpi radialis and pronator teres) is involved

Medial Cord. The ulnar, the medial cutaneous of the arm and the forearm, and the lower half of the median nerve are involved. A combined median and ulnar nerve lesion is produced, except for flexor carpi radialis and pronator teres.

Posterior Cord The radial, the axillary (deltoid, teres minor), the subscapular (subscapularis and teres major) and the thoracodorsal nerves (latissimus dorsi) are involved.

More than one component may be involved partially or completely. Although regression of symptoms occurs in incomplete lesions, an increase in severity and extent may gradually ensue, due to subsequent scar-tissue formation. The cicatrix reaches out and engulfs many originally uninjured portions of the plexus, resulting in a disseminated incomplete motor or sensory disturbance.

Clinical Picture

Usually immediately following trauma, the entire extremity is paralyzed and anesthetic. However, the physiologic portion of injury recovers spontaneously and quickly, leaving the more permanent or slowly improving residuals. Very rarely does one find a clear-cut well-delineated upper or lower plexus syndrome, such as is described later under "Obstetric Paralysis." The study of Erb's and Klumpke's paralysis is useful for localizing the lesion. More frequently, there is an overlap of types or a disseminated irregular involvement.

Treatment

Brachial plexus injuries should be treated immediately and continuously for a long time —months to years. After a brief waiting period for recovery of the temporarily involved muscles, an inventory of the paralyzed muscles is taken, and the lesion is localized If avulsion of the nerve roots is suspected and confirmed by myelography (demonstrated by outpouchings from the dural sac), the situation is hopeless. Fortunately, this condition is rare. Otherwise, early operative intervention is indicated. The poorest results are proportionate to delay in nerve suture. With passage of time, constricting scar tissue at the operative site, muscle atrophy and joint contractures compromise the final result. It is imperative to operate emergently. End-to-end suture of nerves should be the objective. In late cases, surgery is still beneficial. The cicatrix and the neuromas should be excised completely before end-to-end suture is effected Suture may be difficult in old cases with marked retraction, and impossible in avulsion of roots.

Experimentally, homogenous nerve grafts have been used to bridge a gap. It may be necessary to resect the middle third of the clavicle in order to explore the distal plexus. If the 5th or the 6th cervical cannot be sutured, connection to the ansa hypoglossi

may be done. The entire plexus should be explored, by dividing the anterior scalenus and the clavicle if necessary. The exposed nerves are stimulated electrically to determine the injured ones. When a nerve is found in continuity through neuroma formation, it should be left alone. Compression should be relieved. Offending callus or tumor is removed. In aneurysm formation, ligation of the subclavian artery has been done without untoward effects.[5]

Physiotherapy should be instituted immediately after injury and surgery. Avoidance of stretching of paralyzed muscles aids in their recovery. The small and important supraspinatus muscle and deltoid are particularly susceptible. The extremity is placed on an abduction frame in the position of function—90° abduction, 60° external rotation, and 30° forward flexion at the shoulder; the elbow is flexed 90°; the wrist in slight dorsiflexion; and fingers partially flexed with thumb opposed The abduction position and avoidance of the dependent weight of the arm relax the brachial plexus from a constant drag traction Light massage and passive motion are administered, cautiously avoiding overstretching. Galvanic and faradic stimulation directly to the muscles may be beneficial to maintain tone The joints are put through the full range of motion several times daily. As soon as the slightest trace of recovery is detected in a muscle, the motion is encouraged, at first by assistive, later by active, and finally by resistance exercises. It is amazing how even slow, spontaneous recovery can take place by constant, energetic, conscientious physiotherapy in what appears to be a hopeless case. One must remember that the distance from the lesion to the muscle is great; therefore, much time is required before the new nerve fibrils reach their destination Consequently, the end-results should not be determined until after a lapse of several years. However, when it becomes apparent that no recovery can be anticipated in one certain muscle, substitution movements of the other muscles should be encouraged, and reconstructive procedures should be carried out. These include:

Arthrodesis at shoulder at 90° in children, 45° in adults, for loss of abduction

Arthrodesis of elbow at 90° for loss of flexion

Arthrodesis of wrist in dorsiflexion for loss of extension

Tendon transference—wrist flexors to finger extensors and vice versa

Rotation osteotomy of humerus—to correct internal rotation

Steindler advancement operation—when there is failure of active flexion at elbow, and the finger extensors are intact, displacing the point of origin of the common extensor tendon more proximally on the humerus is effective in gaining flexor power.

Many other procedures may be done. If a useless forearm and hand eventuates, it may be preferable to amputate and substitute a clawhand prosthesis.

COSTOCLAVICULAR SYNDROME[6]

When the shoulders are forcibly pulled downward and backward for prolonged periods of time, the space between the clavicle and the first rib is narrowed, the subclavian artery and the brachial plexus are compressed, and neurovascular findings and symptoms are produced. This posture is common in soldiers or others who carry heavy packs on the shoulders. Temporary symptoms of pain and numbness in the arm and the hand and obliteration of the radial pulse contrast sharply with persistence of these findings in the cervical rib and scalenus anticus syndromes. Treatment consists of avoidance of the abnormal posture and performing corrective exercises. Excision of the outer portion of the clavicle may be required.

HYPERABDUCTION SYNDROME[7]

The main vessels and the brachial plexus are subject to stretching and compression at two points. One such site is where they pass beneath the coracoid process posterior to the pectoralis minor. These structures are relaxed in adduction but are stretched about the coracoid in hyperabduction. Strong contrac-

[5] Davis, L , Martin, J , and Perret, P.. The treatment of injuries of the brachial plexus, Ann Surg 125:647, May, 1947.

[6] Falconer, M. A , and Weddell, G · Costoclavicular Compression, Lancet 2·539, 1943.

[7] Beyer, J. A , and Wright, I S · Hyperabduction Syndrome, Circulation 4:161, 1951.

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sensitive vessel may remain in a state of spasm for several minutes after the arm has been adducted.

Treatment requires avoiding the hyperabduction position. Surgery is necessary only if an associated condition such as the costoclavicular syndrome produces symptoms even with minor degrees of movement. Resection of the clavicle or the 1st rib may be necessary. When associated with the scalenus anticus syndrome, scalenotomy is done.

DIFFERENTIAL DIAGNOSIS OF NEUROVASCULAR COMPRESSION SYNDROMES

1. Hyperabduction Syndrome
   - A. *Symptoms and signs of neurovascular disturbance:* Pain (hand, arm and shoulder; shoulder and hand); Raynaud's phenomenon; paresthesias; rubor and/or swelling of hands; weakness; ulceration of the fingertips. Many patients have two or more of these signs or symptoms.
   - B. *Pulse obliterated or neurologic symptoms accentuated in the hyperabduction position*
   - C. *Hyperabduction maintained for prolonged periods during sleep and work.*
   - D. *Cessation of symptoms on adduction.* Recovery may be prolonged if nerve damage is extensive. Edema of extremity may persist if the veins are thrombosed.
2. Costoclavicular Syndrome
   - A. *Symptoms and signs of neurovascular disturbance*
   - B. *Shoulder held downward and backward by traction maintained for prolonged periods*
   - C. *Symptoms reproduced and radial pulse obliterated by forcibly pulling the arm downward and backward*
   - D. *Thoracic outlet narrowed* by cervical rib or deformed first thoracic rib
   - E. *Relief by corrective posture*
3. Cervical Rib
   - A. *X-ray evidence of cervical rib.* A cervical rib in itself does not implicate it as the cause of symptoms. Conversely, its absence does not exclude this possibility, since a fibrous band can produce symptoms.
   - B. *Neurologic findings corresponding to the lower cord of the plexus.* Therefore, the C8 and the T1 dermatomes are involved.
   - C. *Relief following surgical removal*
4. Scalenus Anticus Syndrome
   - A. *Symptoms and signs of neurovascular disturbance*
   - B. *Adson's sign:* Obliteration of the radial pulse and accentuation of neurologic symptoms by forcibly turning the head toward the affected side and taking a deep breath. The test may be reinforced by downward traction on the arm.
   - C. *Hypertrophy or tenderness of the scalenus muscle*
   - D. *Relief by scalenotomy*

CLASSIFICATION OF DISORDERS PRODUCING NEUROVASCULAR SIGNS AND SYMPTOMS IN THE UPPER EXTREMITY:

1. Neurovascular
   - Hyperabduction syndrome
   - Cervical rib syndrome
   - Scalenus anticus syndrome
   - Costoclavicular syndrome
   - Sclerodactylia associated with myocardial ischemia
   - Raynaud's phenomena
   - Scleroderma
   - Neurovascular syndromes due to abnormal first rib, cervicodorsal scoliosis or arthritis, post-traumatic or post-polio defects and poor posture.
2. Predominantly Vascular
   - Raynaud's phenomena
   - Thromboangiitis obliterans
   - Pernio
   - Migratory thrombophlebitis
   - Primary axillary venous thrombosis
   - Diffuse vasculitis
3. Predominantly Neurologic
   - Sympathetic dystrophy
   - Post-traumatic osteoporosis (loosely classified as Sudeck's atrophy)
   - Herpes zoster (postherpetic)
   - Posthemiplegia dystrophies
   - Causalgia
   - Herniated nucleus pulposus (cervical)
   - Peripheral neuritis
   - Syringomyelia

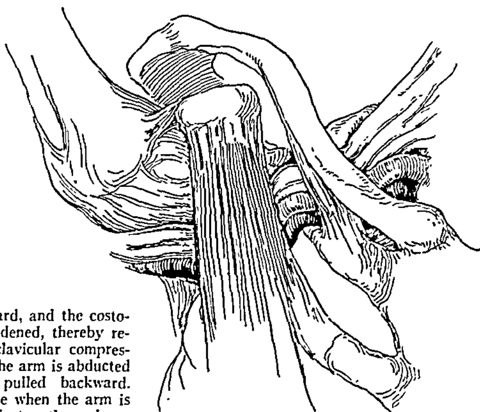


Fig. 302. The hyperabduction syndrome. Extreme hyperabduction stretches the vessels and the nerves beneath the coracoid and its attached tendons. Also shown is the compression of subclavian vessels and brachial plexus *between the clavicle* and the first rib. Actually, at this point, during abduction and external rotation, the arch of the clavicle moves upward, and the costoclavicular interval is widened, thereby relieving pressure. Costoclavicular compression occurs only when the arm is abducted and especially when pulled backward. Obliteration of the pulse when the arm is abducted usually implicates the subcoracoid interval

tion of the pectoralis minor will pull the coracoid downward and similarly stretch the vessels and the nerves. The second point of pressure is between the clavicle and the 1st rib (costoclavicular syndrome). Symptoms due to hyperabduction generally occur because of the position assumed by the patient while asleep. Numbness and paresthesias are noticed first in the fingers and progress centripetally to involve the hands and the arms Pain is not a prominent feature. The pulse is obliterated in the hyperabducted position Occasionally, gangrene of the tips of the fingers may develop. Avoiding the harmful position is curative.

Obliteration of the radial pulse by placing the arm in hyperabduction is very common (83% of normal individuals); therefore, this finding in itself is not diagnostic. In testing for the hyperabduction syndrome, the procedure is as follows: First, the patency of the radial and the ulnar arteries distal to the wrist must be established to rule out occlusive disease as a cause of symptoms. By *Allen's test,* the patient elevates the hand and clenches the fist to express the blood from the hand. By finger pressure the radial and the ulnar arteries are occluded. Then the hand is lowered, and the fist is opened. The arteries are released one at a time: the patency of each one is demonstrated by an immediate flush of blood into the pale skin of the palm. Next, *Wright's test* for the hyperabduction syndrome is performed. The patient is seated with both arms hanging at the sides and the examiner standing behind him. Then the arm is moved passively through an arc of 180°, meanwhile palpating the pulse and noting the point where the pulse diminishes and disappears entirely. Oscillometric readings at various points in the arc define the condition more accurately. Tests to rule out other conditions are performed by (1) traction on the arm downward and backward (test for costoclavicular compression); (2) having the patient turn his head sharply to the right and the left, breathing deeply at the same time, and palpating the pulse or observing oscillometric readings (test for scalenus anticus and cervical rib syndromes). In the hyperabduction syndrome, obliteration of oscillations may take place anywhere in the arc of abduction, and oscillations return promptly upon return of the arm to the side. Occasionally, an unduly

Cord tumors
Crutch palsy

4. **Miscellaneous**
Osteoarthritis, especially cervical
Myofasciitis, primary
Myofasciitis, secondary to osteoarthritis
Dermatomyositis
Nodular panniculitis
Subdeltoid bursitis
Calcium deposits in tendons or other soft tissues

OBSTETRIC PARALYSIS
(Erb's Palsy: Brachial Birth Palsy)

Paralysis of part or all of the upper extremity found at birth generally follows a difficult and prolonged labor. Presumably, the mechanism of production is by forcibly widening the interval between the head and the shoulder, either by laterally flexing the head when a shoulder is caught behind the symphysis pubis or, in breech delivery, by pulling the infant's torso to one side while the aftercoming head is fixed behind the bony rim. Rarely, traction on the arm in wide hyperabduction will be causative. The incidence of the injury has been greatly reduced in recent years because of better obstetric care, particularly by enlarging the indications for cesarean section.

Pathology

The upper portion of the brachial plexus is affected most commonly, usually described as being at the junction of the 5th and the 6th cervical roots (Erb's point). Occasionally, the entire plexus and rarely the lower sections are involved. The lesion may vary from mild stretching with resultant edema and temporary loss of conductivity to actual complete tearing and loss of continuity. The rare lower root injury (C8 to T1) is generally an avulsion proximal to the site of emergence of sympathetic fibers to the first thoracic ganglion; hence, the poor prognosis and the presence of a Horner's syndrome in this type. The typical most common paralysis of the abductors and the external rotators of the shoulder permits the upper end of the humerus to remain in adduction and internal rotation so that the adductors and the internal rotators, mainly the pectoralis major and the subscapularis, and the anterior shoulder joint capsule undergo adaptive shortening. The posterior capsule is stretched and thinned. The humeral head rotates posteriorly and erodes the posterior glenoid rim. A posterior subluxation results. Associated injuries at birth include fracture of the clavicle and upper humerus epiphyseal separation. In consequence of this latter injury which may remain unrecognized, the longitudinal growth of the arm is compromised (fortunately the longitudinal growth factor is greater at the distal end of the humerus), and epiphyseal ossification is delayed and irregular. The acromion tends to enlarge and grow downward, anteriorly and laterally and to form an obstacle to reposition of the humeral head in the glenoid. Paralysis of the elbow flexors (biceps brachialis) allows the upper end of the radius gradually to displace posteriorly over the years. The radial head develops a conical epiphyseal center and becomes lodged behind a flattened capitellum by the age of 8 to 14 The ulna becomes bowed posteriorly.[8]

Clinical Picture

Initially, the newborn infant lies with the extremity lying limp at the side, and typically no muscular activity is demonstrable from shoulder to fingers Swelling may or may not be present and may occur about the clavicle or the shoulder or diffusely from the shoulder to the fingertips.

The infant cries and reacts adversely to attempts to move the extremity passively. Within the first few hours to days, a variable, usually large, amount of muscular activity returns, and swelling and sensitivity to passive movement disappears. Nerve conductivity, temporarily halted by edema fluid, is restored. Typically in Erb's syndrome, the residual paralysis involves the abductors and the external rotators of the shoulder, the flexors of the elbow, and the supinators of the forearm, so that the characteristic "waiter's tip position" results. The arm is in adduction and internal rotation, the elbow is extended, and the forearm is in pronation. When the plexus injury extends a little more distally to C7 components, the extensors of the elbow, the wrist and the proximal phalanges are involved,

[8] Aitken, J. Deformity of the elbow joint as a sequel to Erb's obstetrical paralysis, J. Bone & Joint Surg 34B 352, 1952

so that, *in addition*, the hand displays a flexion attitude. In the rare Klumpke syndrome, involving C8 and T1 components, the intrinsic muscles of the hand are paralyzed, and a clawhand attitude ensues. The medial aspect of the arm and the forearm are anesthetic. Signs of Horner's syndrome may be manifest.

Generally, the course is one of slow, gradual subsidence of the residual paralysis over several weeks to 3 or 4 months, after which further improvement is unlikely. In the Klumpke type the outlook for significant return of function is poor. This is in direct contrast with the Erb type in which prognosis is good, particularly if treatment is instituted early. When the shoulder muscles fail to return adequately, the arm remains fixed in adduction and internal rotation by contractures, chiefly of the pectoralis major and the subscapularis. By *attempting to abduct the arm passively*, the edge of the anterior axillary fold is rendered tense and prominent. The muscles about the scapula and the arm become *atrophic*. The deltoid is thinned. The humeral head hangs dependent away from the acromion and on palpation can be felt to subluxate or even to dislocate posteriorly. A lateral roentgenogram of the glenohumeral joint taken in a transaxillary direction will demonstrate the subluxation and the inadequacy of the posterior glenoid rim. With passage of time, the acromion becomes prominent and curved downward *anterolaterally*. The longitudinal growth of the humerus is slightly retarded. When, secondary to paralysis of the biceps, the radial head is displaced *posteriorly* and deformed, motion of the elbow is restricted by the abnormal articulation. The ulna is bowed posteriorly.

The localized swelling at the proximal areas is frequently due to bony injury. A fractured clavicle is evidenced by crepitus and occasionally palpation of fragment ends. It is difficult to demonstrate an epiphyseal injury very early unless the ossification center is visible. Normally, this lies *in* a superomedial position in relation to the upper end of the shaft. Any deviation suggests an epiphyseal separation. More commonly, proof *of this* injury is forthcoming about 2 weeks later when subperiosteal callus and ossification is demonstrable, encircling the upper end of the shaft. Also, a transverse irregular line of sclerosis becomes apparent within the shaft in the juxta-epiphyseal area and is gradually displaced distally with longitudinal growth. This constitutes mute testimony to the epiphyseal separation. Displacement of the epiphysis laterally and posteriorly predisposes to later development of posterior subluxation.

Differentiation of Paralysis from Bony Injury

Each of these may occur as an independent primary injury and clinically are similar in appearance. Galvanic and faradic currents are used to determine nerve integrity and muscle degeneration if a plexus injury is suspected. Displacement of the ossification center and later development of callus formation and the distally displacing line of sclerosis confirms the diagnosis of fracture through the metaphysis *adjacent to the epiphyseal growth cartilage*.

Treatment

Conservative. Immediately upon establishing the diagnosis the extremity should be placed in a position of 90° abduction and 90° external rotation at the shoulder, 90° flexion at the elbow, full forearm supination and moderate dorsiflexion at the wrist. The arm should not be tied to the head of the bed, as recommended in various texts. This vicious position is invariably one of hyperabduction which stretches the cords of the plexus around the nonyielding coracoid process and further damages the plexus. Instead, pinning the infant down flat on his back on a firm pillow with the arm in the correct position is ideal. It allows for complete infant feeding, bathing *and change of diapers without disturbing the* relationship of the arm to the torso. One should aim at positioning the extremity so that the weakened muscles are relaxed, the tendency to contractures at the shoulder is overcome, and tension on the plexus is avoided. If the rare Klumpke type of intrinsic hand paralysis is present, keeping the extremity fully adducted will protect the lower roots of the plexus. The fingers are splinted with the metacarpophalangeal joints in flexion, the interphalangeal joints in extension, the thumb in slight opposition, and the wrist in moderate *dorsiflexion*.

Daily physiotherapy consists of moving all

joints of the extremity through a full range thrice daily and massaging the muscles very gently in an attempt to preserve their tone. This program is particularly effective as prophylaxis against adduction internal rotation contracture at the shoulder and posterior subluxation of the radius at the elbow.

Although practically no recovery of muscle power occurs after the fourth month, residual muscle weakness demands that splinting and physiotherapy be continued indefinitely until the child attains an age sufficient to co-operate *after surgical corrective measures. Meanwhile,* the use and the strengthening of the recovered muscles is encouraged by intelligent direction of the play of the child, e.g., reaching for objects hanging above the crib or the bed. Any tendency to contractures is counteracted by frequent stretchings.

Completely neglected late cases should undergo a strenuous program of physiotherapy for several years before surgery is undertaken. The scapulothoracic muscles should be strengthened prior to arthrodesing the shoulder. The brachioradialis and wrist extensors undergo intensive and prolonged resistance exercises if the Steindler transposition operation is contemplated.

Surgical. This should be done when the child has reached an age when one can obtain the complete co-operation so necessary to gaining the maximum result of a reconstructive procedure.

SEVER OPERATION FOR INTERNAL ROTATION AND ADDUCTION CONTRACTURE OF THE SHOULDER. The pectoralis major and the subscapularis tendons are severed near their insertion. The tip of the coracoid process with its attached tendons (coracobrachialis, short head of the biceps, and pectoralis minor) is osteotomized completely and allowed to displace downward. If the curved prolongation of the acromion interferes with reduction of a posterior subluxation, removal of the obstructing portion is indicated Then external rotation and abduction should be free and almost normal. In severe late cases, it may be necessary *in addition to tenotomize the latissimus* dorsi and the teres major.

Postoperatively, a cast or a brace maintains the position of abduction and external rotation.

ROTATION OSTEOTOMY. The humerus is osteotomized 2 inches below the joint, the distal fragment is rotated externally 90° and is immobilized for 8 weeks. This procedure is not indicated when posterior torsion or subluxation is present. It does not restore function as effectively as the Sever operation.

CUFF OPERATION FOR POSTERIOR TORSION OR SUBLUXATION OF THE HUMERAL HEAD.[9] The musculotendinous rotator cuff, which envelops the humeral head, is stripped up subperiosteally, the humerus is rotated externally, and the cuff is reattached, the external rotators thereby gaining a new, more anterior position. Aseptic necrosis of the head of the humerus and interference with growth are dangers.

OSTEOTOMY OF SURGICAL NECK FOR POSTERIOR TORSION OR SUBLUXATION. First, the Sever operation is done, thereby correcting the adduction and the internal rotation and reducing the humeral head back into the glenoid. Recurrence of the subluxation inevitably follows when the arm is brought down later. Therefore, after sufficient immobilization in the corrected position, the humeral head is held in its normal relationship to the glenoid while the surgical neck is osteotomized and the distal humerus is rotated internally. Immobilization is continued until the osteotomized site has united. Putti and Scaglietti[10] maintain that many cases of posterior subluxation are in reality primary epiphyseal separation at birth with actual paralysis.

POSTERIOR BONE BLOCK OPERATION. The Sever operation should antedate this procedure by about 6 weeks. A bone graft is driven into the scapula at a point barely within the posterior rim of the glenoid and prevents the backward displacement of the head.

A word of caution is indicated at this time relative to the position of abduction. This should not be maintained uninterruptedly for too long a period of time, else the movement of adduction will be restricted permanently. The use of a brace permits the placing of the arm down at the side at least once daily as a preventative measure.

SHOULDER ARTHRODESIS. When the shoulder

[9] Kleinberg, S: Reattachment of the capsule and external rotators of the shoulder for obstetric paralysis, J A M A 98 294, 1932

[10] Scaglietti, O: The obstetrical shoulder trauma, Surg, Gynec & Obstet. 66 868, 1938

abductors are inadequate, the scapulothoracic muscles, particularly the trapezius, should be strengthened, the Sever operation done, and the shoulder joint fused at 90° in children and 45° in adults.

STEINDLER OPERATION FOR RESTORING FLEXOR POWER TO ELBOW. The common origin of the flexor muscles of the forearm is removed from the medial epicondyle and fixed to a point higher on the humerus. A necessary prerequisite is adequate strength of the flexor muscles.

ARTHRODESIS OF ELBOW. Fixation of the elbow at 90° is the alternative when forearm muscles are inadequate for the Steindler procedure.

ARTHRODESIS OF THE WRIST. Stabilization of the wrist in partial dorsiflexion is necessary for strength of flexor power to the fingers. When the wrist extensors are paralyzed, arthrodesis is probably better than the alternative of tendon transference from the flexors to the extensors.

OPERATIONS FOR INTRINSIC MUSCLE PARALYSIS. This problem is discussed in the section on "The Hand."

21

The Shoulder

SURGICAL ANATOMY

The shoulder is generally thought of as a single ball-and-socket joint—the scapulohumeral articulation However, it consists of 4 separate articulations, the scapulohumeral, the sternoclavicular, the acromioclavicular and the scapulothoracic joints

The scapula is a triangular thin blade of bone which lies in close relationship to the posterior chest wall. It is suspended from the cervical and the thoracic vertebrae by axioscapular muscles (levator scapulae, rhomboids, and upper digitations of the trapezius and the serratus anterior). It is enveloped by muscles, the infraspinati and the supraspinati posteriorly and the subscapularis anteriorly. Bursae in the scapulothoracic interval may become inflamed and cause referred pain in the corresponding extremity. A deformity of the scapula, particularly a forward angulation of the upper medial angle, causes abnormal friction and pain

A triangular projection of bone from the posterior aspect of the scapula, the spine, is the site of attachment of the trapezius posteriorly. At its lateral third, the deltoid attaches. The outer end of the spine, the acromion, overlies the humeral head and also gives origin to the deltoid. The latter muscle anteriorly attaches to the lateral third of the clavicle. The clavicle articulates with the acromion laterally and with the sternum medially

A strong taut ligament, the coraco-acromial ligament, extends from the acromion to the coracoid, a bony projection from the anterior and lateral aspect of the scapula. Coracoid, coraco-acromial ligament and acromion directly overhang the humeral head in close relationship to the latter. Any irregularity or bony hypertrophy of the head will be obstructed during abduction by the coraco-acromial arch, causing friction, pressure necrosis of the soft tissues overlying the head, and pain. The coracoid also gives attachment to the strong coracoclavicular ligaments (trapezoid and conoid ligaments) which firmly hold the clavicle to the scapula.

At the outer upper extremity of the scapula is the small shallow glenoid cavity by which the humeral head articulates with the scapula. A fibrocartilaginous structure, the labrum glenoidale, which is triangular in outline, surrounds the periphery of the cavity. Laterally, it is continuous with the capsule; medially, its sharp border facing the cavity lies free. The long head of the biceps inserts into the supraglenoid tubercle, and its fibers are continuous with those of the labrum. Only a small portion of the humeral head is in contact with the glenoid fossa.

Over the anterolateral aspect of the humeral head, a longitudinally disposed shallow groove, the bicipital sulcus, contains the tendon of the long head of the biceps. It is bordered laterally by the greater tuberosity and medially by the lesser tuberosity. It is covered by the musculotendinous cuff.

The capsule is a fibrous, redundant structure which arises from the capsular border of the labrum and adjacent bone posteriorly and inferiorly. Anteriorly, it attaches to the neck of the scapula at a distance from the glenoid, forming a pouch, the synovial recess. This pocket is conducive to displacement of the head forward. Distally, the capsule inserts beyond the humeral head to the neck and the shaft.

The inner aspect of the capsule is lined by synovium. The synovial lining extends anteriorly into the synovial recess, distally along the bicipital groove where it is reflected to envelop the biceps tendon, and through one or more openings in the capsule to communicate with bursae. Broad, flat tendons of the

rotators (supraspinatus, infraspinatus, teres minor) envelop the head and become incorporated into and strengthen the capsule. This thick covering is known as *the musculotendinous cuff*. The coracohumeral ligament originates from the coracoid process and passes outward and downward in the interval between the supraspinatus and the subscapularis, bridging the bicipital groove, and inserting into both tuberosities. This ligament forms an integral part of the musculotendinous cuff and acts as a checkrein to external rotation. In the frozen shoulder, the ligament undergoes contracture and fixes the shoulder in internal rotation.

The musculotendinous cuff consists of the fibrous capsule and the following attached muscles:

1. **Attaching to the Greater Tuberosity**

   TERES MINOR: *Origin*, axillary border of scapula
   Insertion, inferior facet of greater tuberosity
   Nerve supply, branch of axillary nerve

   INFRASPINATUS: *Origin*, infraspinous portion of scapula
   Insertion, capsule and greater tuberosity
   Nerve supply, suprascapular nerve

   SUPRASPINATUS: *Origin*, supraspinous portion of scapula
   Insertion, capsule and greater tuberosity posterior to the bicipital groove
   Nerve supply, suprascapular nerve

   The supraspinatus lies in close relationship to the overlying coraco-acromial ligamentous and bony arch and the deep aspect of the acromioclavicular joint. An osteo-arthritis of the acromioclavicular joint with bony lipping and enlargement sets up an irritative subacromial tendinitis.

2. **Attaching to the Lesser Tuberosity**

   SUBSCAPULARIS: *Origin*, subscapular fossa
   Insertion, lesser tuberosity, fusing with the capsule over the glenoid rim
   Nerve supply, subscapular nerve

The two constant bursae are the subacromial and the subcoracoid, which frequently communicate with each other and occasionally, through openings in the capsule, with the intraarticular synovium. These bursae separate the supraspinatus from the overlying arch. The subacromial bursal floor is firmly fixed to the greater tuberosity and its roof to the undersurface of the acromion. Lesions of the humeral head, the musculotendinous cuff and the overlying arch and the acromioclavicular joint are often reflected in traumatic inflammation of these bursae. Like bursae elsewhere they may be involved by infection and gouty deposits.

The neurovascular bundle, the axillary nerve with its branch to the teres minor, and the posterior circumflex vessels cross anterior to the subscapularis muscle. This separates these vital structures from the inferior aspect of the glenoid and explains the infrequency of paralysis following antero-inferior dislocation.

At the inferior aspect of the glenoid, the long head of the triceps is attached. Dislocations must occur anterior or inferior to this structure.

The deltoid muscle originates at the outer border of the acromion and the outer thirds of the scapular spine and the clavicle. It is inserted into the deltoid tubercle, a bony prominence at the middle of the humerus on its outer aspect. Its nerve supply is the axillary nerve which winds around the neck of the humerus and enters the muscle posteriorly at about its middle third. Therefore, in surgical exposures of the shoulder, it is essential to avoid traumatizing the muscle beyond 2 inches from its upper attachments.

ARM-TRUNK MECHANISM

The muscles which attach to the scapula, the clavicle and the humerus function as a co-ordinated unit to produce a smooth movement of the shoulder. Motion takes place at the glenohumeral, acromioclavicular, thoracoscapular, sternoclavicular and superohumeral articulations and within the biceps tendon sheath.

When the arm is elevated in abduction or forward flexion, the scapula at first must seek a position of stability, the so-called "setting phase." During the first 30° to 60° of elevation, the scapula moves forward or backward or may even oscillate until it reaches the most stable position. Then it moves laterally with the humerus in the ratio of 1:2. This implies that for every 30° of elevation of the arm the thoracoscapular movement is 10° and glenohumeral movement is 20°. When either of these articulations is fixed by injury or

disease, the loss of motion is proportionate, the glenohumeral fixation causing twice as much restriction as thoracoscapular fixation. Scapulohumeral rhythm is lost. Thoracoscapular movement can effect elevation of the arm to approximately 65° even in the absence of glenohumeral motion. This is the so-called "shrugging mechanism."

Stability of the scapula is accomplished by the co-ordinated contractions of the following muscles attaching to the scapula:

Deltoid. Paralysis prevents abduction beyond 30°. Further elevation of the arm is accomplished by the shrugging mechanism.

Trapezius. It is innervated by the spinal accessory nerve. It acts to rotate the scapula in a clockwise direction. Paralysis allows the acromion and the glenoid to drop to a lower level, the superior angle to lie at a higher level, and the vertebral border is displaced away from the mid-line.

Rhomboidei. These act to carry the inferior angle medially and upward, effecting a counterclockwise rotation of the scapula. Its paralysis results in the scapula displacing away from the mid-line in a clockwise rotation.

Levator Scapulae. Its action is to raise the medial angle of the scapula. Its weakness allows the medial border of the scapula to descend over an abnormal distance as the outer end of the scapula is elevated.

Serratus Anterior. This carries the scapula forward, particularly in the act of pushing. When it is paralyzed, the vertebral border protrudes backward (winging) and is displaced backward toward the vertebral column during abduction.

Pectoralis Minor. This depresses the point of the shoulder downward and medialward. Its loss results in abnormal elevation of the scapula by the trapezius during abduction.

When scapular stability is impaired by weakness of these muscles, elevation of the arm above the horizontal plane is difficult. However, uninvolved muscles eventually compensate so that elevation of the arm improves somewhat, although some instability always remains. When the scapula becomes fixed as after thoracic operations, abduction of the arm above the horizontal plane is impossible. With the arm held at 90° of abduction, forward and backward movement is limited markedly.

Claviculohumeral Mechanism

During the first 90° of arm abduction, the clavicle at the sternoclavicular joint elevates 4° for every 10° of elevation of the arm. Beyond 90° movement at this joint is nil. At the acromioclavicular joint, motion takes place during the first 30° of abduction and after 100°. The total movement at this joint is only 20°. During abduction, the clavicle rotates on its long axis. If the clavicle is prevented from rotating by fixation at the outer or the inner ends, abduction of the arm above 110° is prevented. Therefore, it is unwise to arthrodese the acromioclavicular joint.

Stability of the Humeral Head

The supraspinatus, the teres minor and the infraspinatus are attached to the greater tuberosity and act to rotate the humerus externally. The subscapularis, which attaches to the lesser tuberosity, is the internal rotator of the humeral head. Together these muscles completely envelop the head and co-ordinate to fix the head in the glenoid and cause the head to descend as the humerus is abducted. The latissimus dorsi and the pectoralis major which attach to the margins of the bicipital groove in addition to their function of adducting the arm aid in depressing the upper end of the arm during abduction.

The tendon of the long head of the biceps originates at the supraglenoid tubercle on the superior rim of the glenoid fossa and passes through the bicipital groove beneath a fibrous sheath extending between the subscapularis and the supraspinatus tendons. The tendon glides passively through the tunnel when the arm is moved. It moves downward on abduction, especially with the arm in internal rotation and forward flexion. Upward movement occurs in adduction, especially with external rotation and backward flexion. In the act of abduction, depression of the head of the humerus is essential. Contraction of the biceps tightens the tendon of the long head over the humeral head, the latter being pushed downward away from the acromion. When the biceps muscle is weak, or when the tendon is fixed to the groove, impingement of the greater tuberosity against the acromion results in a shrugging type of abduction. Eventually, the intra-articular portion of the tendon is worn

away, the bony prominence lessens from erosion, and the acromiohumeral interval again becomes adequate, and full range of abduction is restored.[1]

EXAMINATION OF THE SHOULDER

History

An accurate history of trauma is elicited; of onset, whether acute or chronic; type of pain and area of referral; and the specific kind of disability noted by the patient.

General Examination

The general body habitus is noted. Poor posture denotes the fatigue state which causes pain in the trapezius area and other muscles of the shoulder girdle. Possible causes of pain in the upper extremity are sought in the cervical spine, the chest and the subdiaphragmatic region.

Local Examination

The general contour of the shoulder is observed to detect atrophy. Reflex sympathetic dystrophy is evidenced by swelling, cyanosis, shiny skin, excessive or diminished sweating in the palm, etc.

Active motion is studied in abduction; in adduction, including the ability to bring the arm across the chest toward the opposite shoulder; in external rotation, as in reaching behind the head; in internal rotation, as when reaching behind between the shoulder blades; and forward and backward flexion. The affected extremity should be compared with the opposite extremity. Limitation of motion is recorded for future comparison. When abduction is limited, the patient tends to shrug the shoulder in an attempt to elevate the arm by scapular motion.

Next, passive motion is tested. The examiner fixes the scapula by holding the shoulder down, and glenohumeral passive motion is estimated, recorded and compared with the range of active motion. If passive motion is normal as compared with restricted active movement, the deficient motion is due either to the patient's unwillingness to raise the arm to the painful point or to muscle weakness. When the rotator cuff is ruptured, abduction is greatly limited, but the arm can be raised with assistance. However, active abduction cannot be sustained against resistance, and the arm falls quickly to the side. Then the cuff is palpated while the arm is elevated with the extremity in different degrees of rotation. This will define accurately the site of the tear.

As the injured area of the tendon strikes the acromion, an acute pain will be experienced. At the same time a sense of crepitus or a soft snap will be detected. The point of tenderness localizes the lesion, whether it be a tear or a calcium deposit. This is usually located above and proximal to the greater tuberosity. The anterior aspect of the head is examined while the arm is flexed backward. Forward flexion of the arm renders the posterior region of the cuff accessible to palpation below the acromion. The acromioclavicular joint should be examined for crepitus and tenderness as a possible site of degenerative arthritis, an indirect cause of underlying bursitis and cuff lesions. When this joint is involved, it is manifest by the inability to adduct the arm across the chest. Attempts to perform this motion are painful. Inflammation of the bicipital sheath is evidenced by tenderness along the groove. If the tendon has been torn from its groove, the Yergason sign is positive. This is performed by having the patient actively flex the elbow against resistance and at the same time rotate the arm externally. Subluxation of the tendon is demonstrated by a painful snap about the bicipital groove. The muscles about the scapula should be examined for tender fibrous nodules as a source of shoulder and arm pain.

Procaine Infiltration

If a lesion is suspected in the musculotendinous cuff, injection of a local anesthetic into this area will eliminate the pain. The needling of a tender area, whether it be due to calcium deposit or a minor cuff tear, is an excellent therapeutic measure; it should be repeated at intervals.

Roentgenologic Examination

The shoulder is examined by anteroposterior views with the arm in internal and external rotation. By reducing the kilovoltage, the faint shadows of calcification may be

[1] Turek, S L: The painful and stiff shoulder, J. Internat Coll. Surgeons **22** 695, 1954.

brought out. A lateral view of the humeral head is made by a transthoracic exposure or by the transaxillary view. The cervical spine is roentgenographed routinely to rule out the presence of a cervical rib or a protruded cervical disk.

LESIONS OF THE ROTATOR CUFF

Etiology

The tendinous fibers of the rotator muscles at or near their insertion into the tuberosities undergo degenerative changes with advancing age. Deterioration is pronounced after the fifth decade and is observed in all shoulders after the age of 60. The point of attachment where degenerative changes are most severe is known as the "critical area" This is the same site where calcified deposits are seen Disintegration of the tendon predisposes to rupture, so that at the sixth decade defects in the cuff are seen almost universally. Before the fifth decade, the tendon is strong, and avulsion of the tuberosity is more probable. The tendon is worn down by attrition between the humeral head and the coraco-acromial arch. The biceps tendon over the humeral head is likewise eroded and is predisposed to rupture. As the tendon disintegrates and disappears, the remainder of the tendon becomes fixed in the bicipital groove. As the overlying soft tissues are worn down, the tuberosity is subjected to pressure and friction as it rides against the undersurface of the acromion, and it too becomes eroded. The bone at this site becomes sclerotic and cystic. As the cuff wears away, the deltoid gradually takes over more function. *The syndrome* is manifest clinically by recurring pains and stiffness, aggravated by activity, occurring in the shoulder and radiating down the anterior aspect of the arm. Tenderness is perceived over the cuff about the greater tuberosity and over the bicipital groove

Clinical Picture of Acute Rupture

A history is often obtained of a fall on the shoulder or an attempt at lifting a heavy object outward. A severe acute pain and snap in the shoulder is felt immediately, and the patient is unable to abduct the arm The pain becomes progressively severe over the next few hours. In the senile individual in whom the tendon is quite degenerated, rupture may occur with trivial trauma and a minimum of pain. Often the pain is referred to the deltoid insertion.

Examination

The arm can be raised actively to about 45° by the deltoid, after which elevation of the arm is accomplished by the shrugging mechanism. The pain is most severe in the arc from 45° to 90° when the traumatized tendon is compressed between the tuberosity and the coraco-acromial arch.

If the arm is elevated passively above the horizontal, further abduction is possible with very little discomfort. The pain is most pronounced when the painful tear is exposed to compression.

If the tear is in the posterosuperior portion, pain in the arm is most severe when the arm is rotated internally during abduction. If the tear is in the anterosuperior aspect, pain is excruciating when the arm is abducted and rotated externally.

The strength of abduction is proportionate to the size of the tear. By injecting the site of maximum tenderness with local anesthetic, pain is relieved, and then examination for active abduction can be conducted. If the tear is extensive, abduction is weak and is performed through a small range. If the tear is small, the range of motion may be complete. However, when resistance is offered to abduction, weakness is manifest by the arm dropping to the side.

The anterior aspect of the cuff may be palpated if the arm is extended backward. The posterior aspect is more perceptible to palpation if the arm is forward flexed. A stub of tendon or an actual gap in the deep soft tissues may be felt adjacent to the bony prominence of the greater tuberosity. As the arm is abducted, rough grating is palpable at this point, and crepitus may be audible Similar findings are noted adjacent to the lesser tuberosity if the subscapularis is involved. If the biceps tendon is ruptured, the muscle belly is seen and felt to be shortened and rolled up into a mass in the distal third of the arm Elbow flexion is weak or impossible

Subsequent Course

The pain gradually lessens over a number of months, and the ability to abduct improves

Eventually, a full range of painless motion results. However, the arm is weak for more than ordinary use and useless for sustained work above the horizontal. Atrophy of the supraspinatus and the infraspinatus renders the scapular spine prominent. The abductor and internal rotator muscles undergo contracture unless counteracted by continuous exercises.

Roentgenologic Findings

In the early acute case the roentgenogram is negative. Eventually, over the ensuing months, the tuberosity recedes and becomes irregular, sclerotic and cystic.

Treatment

When preservation of strength for physical activity is desirable, the tendon should be repaired immediately. Through a transacromial approach, the edges of the tear are approximated and sutured. Postoperatively, the arm is placed in a cast in a position of 90° abduction, 60° external rotation and 30° forward flexion. Immobilization for 3 weeks is necessary for healing, after which active exercises are instituted. If repair is delayed, the edges of the tear retract, and approximation is difficult. Contraindications to repair are old age and sedentary occupations.

Occasionally, a contusion of the cuff, a slight tear, or a traumatic bursitis may simulate a complete tear clinically. In such a doubtful case, a period of watchful waiting is acceptable, during which time the arm is immobilized in the "90-60-30 position."

A neglected case during the period of inactivity will develop stiffness and marked limitation of motion of the shoulder. Physiotherapy, including heat, stretching and active and passive exercises, will mobilize the joint sufficiently for better evaluation When full passive abduction is possible, active abduction by the shrugging mechanism defines a tear of the cuff Repair is indicated

Patterns of Rupture

The supraspinatus is most commonly ruptured and at a right angle to the direction of its fibers near its point of insertion into the greater tuberosity. This transverse tear may extend proximally at a right angle and from one corner of the original tear. Usually, the proximal extension occurs between the supraspinatus and the subscapularis. The tear may extend distally through the infraspinatus and the teres minor. When the subscapularis is torn, the biceps tendon is avulsed from its groove and displaced medially. When both supraspinatus and subscapularis are torn, a massive avulsion of the cuff results. Occasionally, the biceps tendon is torn as an associated injury. Rupture of the subscapularis tendon does not interfere with abduction and is rarely diagnosed. It is a frequent concomitant injury in dislocations of the shoulder.

Technic of Repair of Acute Ruptures

Under inhalation anesthesia, either in the sitting or the recumbent position with a sandbag under the shoulder, a longitudinal muscle-splitting incision is made anteriorly in the deltoid, extending from the acromion inferiorly for a distance of no more than 2 inches (to avoid the axillary nerve). An incision is made in the bursal roof. The bursal floor is attached intimately to the underlying cuff. Rotation of the arm reveals the defect. In the case of the supraspinatus, 90° abduction, 60° of external rotation and 30° of forward flexion usually approximate the edges of the tear. The distal stub of the tendon and the underlying compact bone are removed, forming a slot into the cancellous bone. Mattress sutures of No. 2 braided silk are used for tension sutures, i.e., in that part of the tear which is at a right angle to the direction of the fibers. They are brought through drill holes in the bone. Interrupted sutures of heavy silk approximate the longitudinal tear. In cases of advanced degenerative periarthritis where the cuff is thinned, repair may be impossible. Regardless of that condition, the biceps tendon should be fixed firmly in its groove, thereby aiding stabilization and reducing the possibility of later rupture. Following repair, tension on the suture site is lessened, and healing aided by the 90-60-30 position.

In order to avoid compression of the repaired tendon against the coraco-acromial arch, the acromion and the coraco-acromial ligament may be removed without subsequent functional loss.[2]

[2] [illegible] G. Lesions of [illegible] der, J. Bone & [illegible]



FIG. 303. Calcified tendinitis. The true extent of the deposit is revealed by underexposed roentgenograms and the proper rotational position of the humerus.

Repair of Chronic Ruptures of the Cuff

At operation one finds a triangular or half-moon-shaped defect with the base at the tuberosity, with smooth rounded edges and the proximal portion retracted beneath the acromion. When the long head of the biceps displays congestion and thickening of its synovial sheath, the intra-articular portion likewise is irritated. The tendon may be frayed and even worn away. An avulsed greater tuberosity may heal in a malunited position, protruding through a rent in the cuff, the edges of which are formed by the supraspinatus and the subscapularis. When the biceps tendon has been avulsed from the groove with the subscapularis tear and displaced medially, the intra-articular portion should be removed. If in addition a dislocation has occurred, a Platt procedure should be done. The edges of the cuff tear are freshened, and sufficient bone is resected from the tuberosity to allow the humeral head to pass unobstructed beneath the acromion. As an alternative, the acromion may be resected, providing easier access to the tear. The arm is placed in the 90-60-30 position, and an attempt is made to suture the tear. However, retraction often makes repair impossible. Therefore, side-to-side approximation by interrupted sutures of heavy silk is done. Distally, tension mattress sutures are inserted, usually at a more proximal position in the head. This necessitates creating a slot in the cartilage-covered portion of the head. The obvious result is some loss of abduction range.

Postoperative Care

The arm is placed in a cast in the 90-60-30 position for 5 or 6 weeks. Thereafter the arm is gradually brought down to the side, over several weeks, to avoid undue tension and reinjury of the tendon. At first, abduction exercises without gravity are practiced; later, against gravity. Next, pulley exercises are done, at first in forward and backward flexion; later, in abduction. After a full range of active motion has been obtained, resistance exercises are prescribed. At least 6 months is required for maximal recovery.

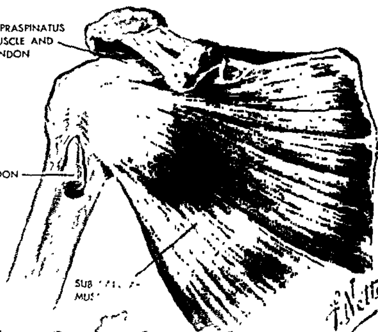
CALCIFIED DEPOSITS IN THE ROTATOR CUFF

The deposit of amorphous calcium phosphate and calcium carbonate in the rotator cuff is a very frequent occurrence. These salts deposit in tendons, ligaments, aponeuroses and capsular attachments as well as blood vessel walls. This deposit is associated with degenerative changes, Codman described a so-called "critical zone" near the attachment of the musculotendinous cuff to the tuberosity, where degenerative changes allow a rupture to take place, and, on the other hand, cause a necrosis with subsequent deposits of calcium in this area. The deposits are found initially in the tendon and work their way to the surface, eventually rupturing into the bursa. While it is under tension within the tendon, it irritates the overlying bursa, which exhibits a reddened, congested appearance in the area immediately overlying the deposit. Within the center of the reddened area, the yellowish or whitish appearance of the calcium deposit shines through. When the condition is chronic, the overlying area has a bluish discoloration, is thickened, and multiple adhesions or villous formations may occur. This latter condition is an adhesive or villous chronic subdeltoid bursitis which is secondary to the chronic irritation of the calcific deposit. In the chronic stage, the deposit is thick and resembles tooth paste in consistency and becomes liquid in the acute stage. After bursting into the bursa, the material is absorbed, and the condition subsides spontaneously. Recurrent attacks of calcific deposits with irritation of the overlying bursa result in a chronic adhesive bursitis.

Calcific tendinitis occurs chiefly in the white-collar class. It is frequently bilateral,

SUBACROMIAL BURSA
SUBCORACOID BURSA
CORACO-ACROMIAL LIGAMENT
SUPRASPINATUS MUSCLE
SUPRASCAPULAR NERVE AND ARTERY
CORACOID PROCESS
ACROMION PROCESS
CIRCUMFLEX (AXILLARY) NERVE
LONG TENDON OF BICEPS
BRANCHES OF AXILLARY NERVE
DELTOID MUSCLE
MUSCULO-CUTANEOUS NERVE
SUBSCAPULARIS MUSCLE
RADIAL NERVE
MEDIAN NERVE
ULNAR NERVE

NEURO-VASCULAR RELATIONS OF THE SHOULDER JOINT



SHOULDER JOINT FROM IN FRONT WITH BURSAE AND OTHER OVERLYING STRUCTURES REMOVED

DELTOID

TRAPEZIUS

C7

T2

T12

LEVATOR SCAPULAE

RHOMBOIDEUS MINOR

RHOMBOIDEUS MAJOR

SUPRASPINATUS

INFRASPINATUS

TERES MINOR

ACROMION PROCESS

SUBACROMIAL BURSA

REFLECTED DELTOID

CIRCUMFLEX (AXILLARY) NERVE

TERES MAJOR

LONG HEAD OF TRICEPS

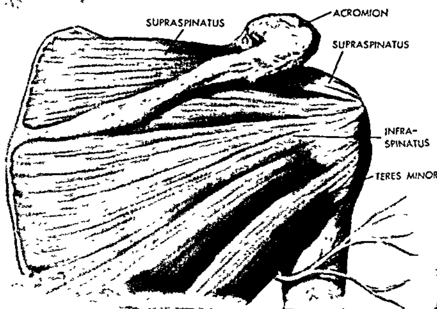
LATERAL HEAD OF TRICEPS

LATISSIMUS DORSI

LATISSIMUS DORSI

THE SHOULDER DISSECTED FROM THE REAR

DETAIL WITH BURSA REMOVED TO SHOW INSERTION OF SUPRA- AND INFRA-SPINATI IN FORMATION OF ROTATOR CUFF



F. Netter M.D

PLATE 44. (From Moseley, H. F., and Lampe, E. W.: C" ... posia. ... F. Netter)

RUPTURES OF THE ROTATOR CUFF

RUPTURES OF SUPRA- AND INFRA-SPINATI

(Humerus rotated medially to expose rupture lateral to biceps tendon)

REPAIR

ACUTE RUPTURE

CHRONIC RUPTURE

ACUTE SUPERIMPOSED ON CHRONIC RUPTURE

RUPTURES OF SUBSCAPULARIS TENDON

(Humerus rotated laterally to expose rupture medial to biceps tendon)

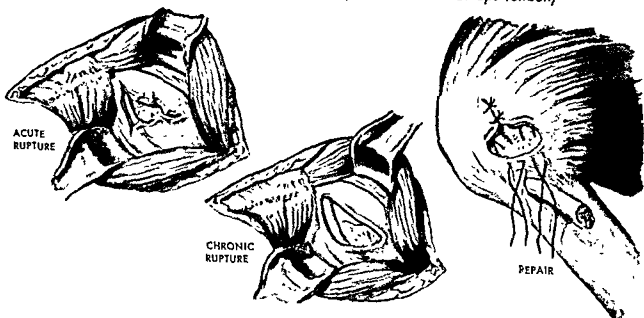
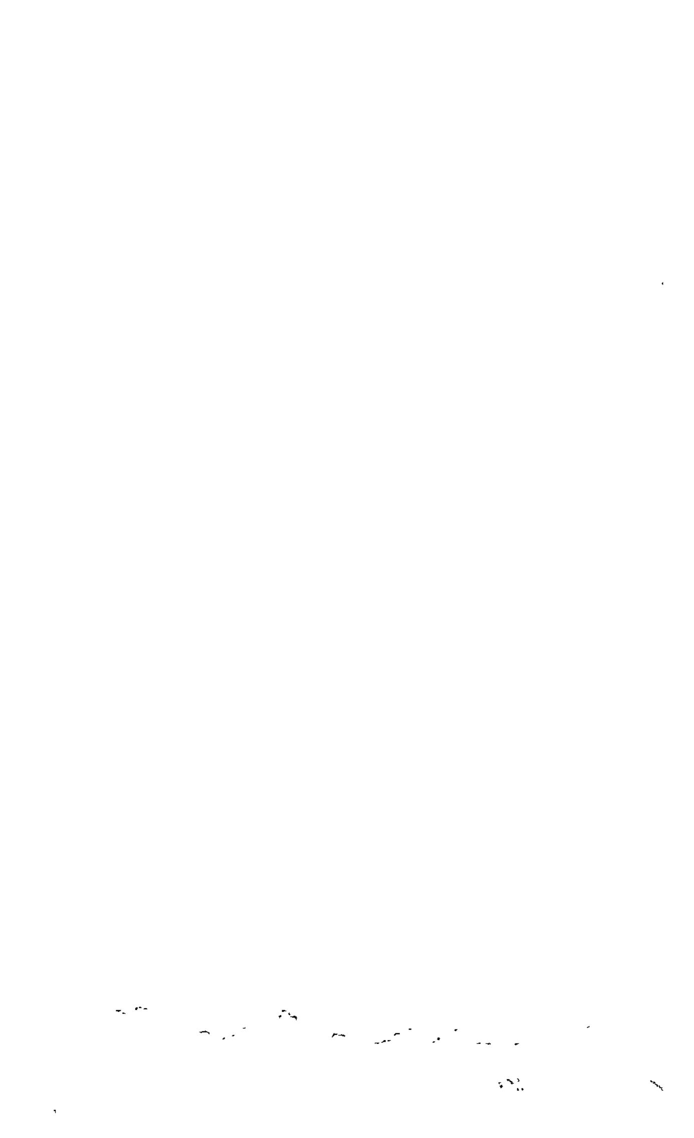


PLATE 45. (From Moseley, H. F., and Lampe, E. W.: Ciba Clinical Symposia. Drawing by F. Netter)



and often there is an associated history of rheumatic pain in the shoulders and other joints. Not infrequently cardiovascular disease is associated. Males in the third and the fourth decades are predisposed, although females and other ages are affected.

Clinical Picture

The onset may be gradual or very acute and may follow an injury to the shoulder such as a sprain or overuse. Over a period of 2 or 3 days, the acute pain develops about the humeral head and becomes quite intense. The pain may intensify over the next few days and then gradually diminish, indicating that the tension of the deposit has diminished or that the calcium has ruptured into the bursa, the bicipital groove, or the adjacent fascial plane. During the painful period, limitation of abduction is noted. The patient cannot bring the arm to the point where the tender area impinges against the acromion. However, with the shoulder relaxed so that the humeral drops away from the coraco-acromial arch, the arm can be elevated passively with a minimum of pain. Pressure upon the painful area, as when lying on the affected shoulder, is painful. Tenderness over the deposit is excruciating. A roentgenogram taken with reduced kilovoltage with the arm in internal and external rotation will bring out the true extent of the deposit. A transaxillary view will reveal a deposit located in the subscapularis, the infraspinatus or the teres minor.

Treatment

Conservative. An icebag to the shoulder reduces the congestion and effectively lessens the pain. The patient rests in bed in the sitting position so that the weight of the arm in dependency removes the compression of the bursa and the tendon between the acromion and the humeral head. Sedatives are given. Under local anesthesia, a large-bore needle is inserted into the area of the deposit, and multiple punctures allow the deposits to exude into the overlying bursa. If one wishes to remove the material, a second large-bore needle may be inserted into the bursa about ½ inch away from the original needle. Through-and-through irrigation with normal saline will wash out flakes of calcific material. Dramatic relief may be obtained. A slight soreness remains a few days after needling and then quickly subsides. Following the needling, hot packs are administered, and exercises are started to prevent shoulder stiffness. Gentle massage and short-wave diathermy are added. Pulley exercises are started as soon as they can be tolerated. By this treatment, recurrences are infrequent.

Recently, ACTH and cortisone and their derivatives have been used with excellent results. X-ray therapy has been used with questionable results.

Surgical. Codman, Bosworth, Moseley and others have shown that operative treatment relieves the acute pain completely and permanently. Recurrences are rare. Moseley's criteria for surgical removal are: (1) large deposits in the acute stage; (2) large deposits and recurrences resistant to conservative therapy; (3) large deposits which are likely to impinge upon the coraco-acromial arch.

Technic. Under general anesthesia, a transverse incision is made. The deltoid is split. The roof and the floor of the bursa are removed, and the calcific deposit is located by rotating the arm. All material is curetted out. If too large a defect remains in the tendon, it should be closed with strong silk sutures. A compression dressing is applied. The subsequent treatment is similar to that following the needling procedure.

BICEPS TENDON LESIONS

Anatomy

The tendon of the long head of the biceps arises from the tubercle on the superior rim of the glenoid fossa and passes in close approximation to the articular surface of the humeral head toward the bicipital groove. Where it enters the groove it is covered by the transverse humeral ligament which extends between the tuberosities. It becomes enveloped by a prolongation of the synovium which, with the tendon, enters the bicipital groove. At the distal end of the groove, the synovium is reflected backward to cover the extra-articular portion of the tendon. The groove is covered over by fascial extensions of the subscapularis tendon at the upper end and of the pectoralis major at the lower end. The intra-articular portion of the tendon, before it enters the

groove, is in close relationship superficially to the capsule and the coracohumeral ligament, the latter lying between the subscapularis medially and the supraspinatus laterally. When movement occurs at the glenohumeral joint, the tendon glides within the groove. On abduction and forward flexion, the tendon glides distally. On adduction, backward flexion and external rotation, the tendon glides proximally into the joint.

Tenosynovitis of the Biceps Tendon

Inflammation of the surrounding synovial sheath is a result of (1) *trauma* (contusion, tear of the cuff, tear of subscapularis insertion which avulses the biceps from the groove, tendinitis of the cuff); (2) *calcific deposits* in the cuff; and (3) *infection,* which is usually an extension from intra-articular involvement. Bicipital tenosynovitis causes pain over the anterolateral aspect of the shoulder referred along the anterior surface of the arm. The pain is aggravated by shoulder motion, is worse at night and is relieved by rest. Protective muscle spasm of the deltoid and the scapular muscles is noted in the acute stage. Active and passive movements are restricted by the patient's reluctance to accentuate the discomfort. The following tests aid in making the diagnosis:

1. Forced external rotation of the arm reproduces the pain over the bicipital groove.
2. Faradic stimulation of the biceps tendon produces the same pain.
3. By grasping the tendon above its attachment to the muscle belly and moving it from side to side, pain over the groove is increased
4. Yergason's sign. This is performed by flexing the elbow and having the patient forcefully supinate the forearm against resistance, the resulting forceful contraction of the biceps effects distal movement of the tendon and causes pain in the bicipital groove. These tests are frequently positive in older patients with degenerative changes about the humeral head. The irregularities of bone about the groove cause friction and irritation of the tendon as it glides.

Treatment. In the acute stage, the shoulder is placed at complete rest until the pain subsides. Then active gentle motion is started, and usually a full range is restored. However, if the tenosynovitis has lasted a long time, adhesions will form about the tendon within the groove and in the intra-articular portion which will adhere to the capsule. On attempted abduction, the tendon will not glide, and the intra-articular segment will coil up and obstruct movement. Motion of the joint is restricted from 6 months to several years. Eventually, the intra-articular tendon undergoes attritional changes and absorbs, the extra-articular tendon becomes firmly fixed in the groove, and motion is restored. Therefore, if after the acute stage motion does not return quickly, the intra-articular tendon may be resected to effect rapid return of function. Adherence of the tendon rarely occurs in individuals before the age of 40.

Lippmann and DePalma state that tenosynovitis of the long head of the biceps is the main factor in restricting motion in the frozen shoulder. Lippmann removes the extra-articular tendon and transplants it to the coracoid process.[3] DePalma, in addition, removes the intra-articular portion which he says restricts motion by its adherence to the capsule.

Infections are treated by antibiotics, rest, hot packs and fluids. After signs of inflammation have subsided, further treatment is similar to that used for the noninfectious type.

Attrition and Rupture of the Biceps Tendon

With advancing age, the tendon may become degenerate and soft. Degenerative changes of bone cause excrescences about the intertubercular sulcus and roughening of the margins of the bicipital groove. Constant friction against a degenerate tendon causes shredding and fraying, and the tendon becomes thinned and lacks resistance to tension forces exerted upon it by the muscle. As a result, the tendon elongates, and the biceps becomes inefficient. Normally, when the elbow is flexed and the forearm is supinated against resistance, the belly of the biceps is firm, and its tendon cannot be palpated. However, when the tendon is elongated, the muscle belly is not firm, and the tendon can be perceived as a rounded structure in front of the deltoid insertion.

[3] Lippmann, R. K.: The frozen shoulder, S. Clin. North America 31:367-383, 1951.

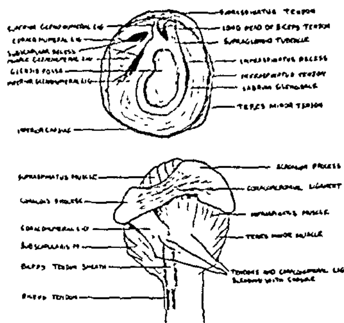


FIG. 304. (*Top*) Glenoid aspect of shoulder joint. Note multiple plications and redundancy of capsule and its intimate relation with tendons of rotator muscles. Bicipital tendon attaches to the cartilaginous labrum and upper bony rim. (*Bottom*) Anterolateral view of subdeltoid structures. Note close approximation of coracoacromial ligament to the subjacent capsule. (Turek, S. L.: The painful and stiff shoulder, J. Internat. Coll. Surgeons 22:695)

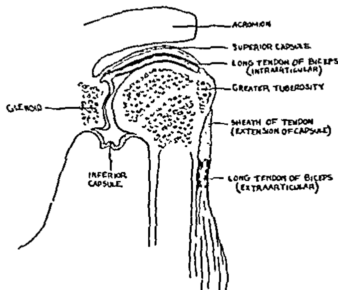


FIG 305. Frontal section through shoulder joint. Acromion, superior capsule, biceps tendon and humeral head are actually in close contact in resting position but are shown as separated structures for clarity. (Turek, S. L.: The painful and stiff shoulder, J. Internat. Coll. Surgeons 22:695)

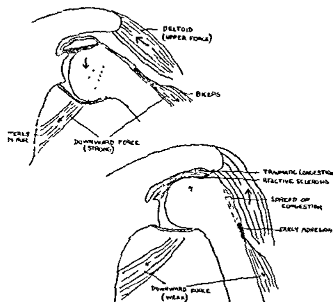


FIG 306. (*Top*) Normal mechanics of abduction at glenohumeral joint. Size of arrows indicates relative strength of force. Sum total of downward force of infraspinous muscles and long head of biceps and upward outward deltoid force is a strong downward thrust of the humeral head. (*Bottom*) Abnormal mechanics leading to chronic capsular trauma Depressor effect of biceps and infraspinous muscles is minimal or nonexistent. Result is an upward thrust of the humeral head as deltoid contracts. Repeated compression of capsule about greater tuberosity leads to capsular congestion, bony sclerosis, and recessional changes in tuberosity. (Turek, S. L.: The painful and stiff shoulder, J Internat. Coll. Surgeons 22:695)

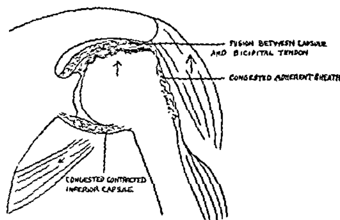


Fig. 307. Stage of severe contraction. Superior capsule and bicipital tendon are swollen and adherent to humeral head and groove. Inferior capsule is thickened and contracted. Deltoid power cannot overcome obstructing subacromial tissues and tight inferior capsule. (Turek, S. L.: *The painful and stiff shoulder*, J. Internat. Coll. Surgeons 22:695)

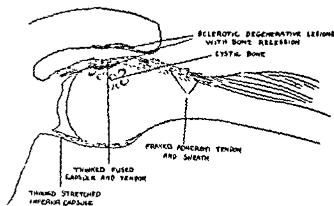


Fig. 308 Final stage. Repeated attempts at abduction have worn down the soft tissues and caused sclerotic and cystic changes in the adjacent bone. The inferior capsule has stretched. (Turek, S L.: The painful and stiff shoulder, J. Internat. Coll. Surgeons 22:695)

The continuous attrition of the tendon within the irregular groove and by compression between the humeral head and the coracoacromial arch thins the tendon until it ruptures. Interruption in continuity may occur spontaneously or during a lifting strain. The biceps muscle is displaced distally in the arm, where it is observed as an abnormally large mass. Rupture of the tendon is accompanied by a sudden sharp pain and immediate weakness in flexing the elbow.

Treatment. If the tendon is elongated but unruptured, the intra-articular portion is removed, and the extra-articular tendon is transplanted to the coracoid process When rupture has occurred, the tendon is found coiled up about the upper end of the muscle belly in the distal third of the arm A tear of the rotator cuff is often associated. Therefore, the shoulder joint is explored, the intra-articular tendon is removed, the cuff repaired, and the distal tendon fixed under tension to the bicipital groove or coracoid. Postoperatively, the elbow is immobilized in flexion for 6 weeks. Thereafter, gradual flexion exercises for the elbow are prescribed.

Subluxation and Dislocation of the Biceps Tendon

Usually, some rotatory injury of the shoulder, such as forcefully rotating the arm externally, is followed by symptoms of painful clicking or thumping in the shoulder with certain movements, particularly external rotation. Often the arm may be locked temporarily in external rotation. By forcefully rotating the arm internally, the tendon slips back into the groove, and the shoulder is unlocked. A click or a thump is palpable over the groove as the arm is rotated externally The pathology consists of the subscapularis tendon being torn away from its mooring, unroofing the bicipital groove, displacing the biceps tendon. Often a prolongation of fascial tissue from the subscapularis envelops the biceps tendon and acts as a sling which elevates the tendon from the groove The tendon can be made to displace from the groove by rotating the humerus externally.

Treatment. Operative exposure is made through the coracohumeral ligament, which lies between the supraspinatus and the subscapularis portions of the cuff. The intra-articular tendon is removed, and the distal tendon is fixed to the groove. The subscapularis tendon is repaired at the same time.

THE FROZEN SHOULDER

The frozen shoulder is a condition of unknown etiology characterized by a gradually progressive, painful, restriction of all joint motion, chronicity, and slow spontaneous restoration of partial or complete motion over months to years.

Clinical Picture

Individuals in the fifth and the sixth decades, particularly women with cardiovascular disease, are predisposed. The onset is insidious and develops during a period of relative inactivity in use of the shoulder, the arm hanging constantly at the side. Some vague antecedent injury may be blamed for the increasing pain and stiffness in the shoulder. The pain is located over the anterolateral aspect of the joint and radiates to the anterior aspect of the upper arm and occasionally to the flexor aspect of the forearm. Discomfort is worse at night and interferes with sleep. Tenderness is generalized about the humeral head and over the bicipital groove. Active and passive motion is limited in all directions, pain being accentuated at the extremes of motion. Early, in the acute stage, muscle spasm is observed in all muscles about the shoulder, particularly the pectoralis major and the scapular muscles. The arm is held protectively at the side in a position of internal rotation. As the condition progresses over several weeks or months, motion gradually diminishes, and pain lessens. Eventually, little or no motion remains, and pain is slight or absent. After subsidence of pain, there begins a slow restoration of movement in the joint. This takes place over an indefinite period varying from months to years. The final status of the shoulder may range from moderate limitation to full restoration of motion. In most cases all but about 10° or 20° of full movement returns. During the period of fixation and nonuse the scapular muscles become atrophic, so that the scapular spine becomes prominent, the humeral head is held high against the acromion as compared with the opposite side, and the pectoralis major is contracted. Often signs of reflex sympathetic dystrophy develop in the hand: swelling of the fingers, shiny atrophic skin, mottled dusky discoloration, coldness, hyperhydrosis or hypohydrosis, hypersensitivity, marked limitation of motion, etc.

Etiology

The actual cause is unknown. The following have been blamed:

1. Tendinitis of rotator cuff (Codman) and other shoulder injuries
2. Bicipital tenosynovitis (Pasteur, Lippmann, Hitchcock and Bechtol)
3. Muscle imbalance, developing from inactivity
4. Reflex sympathetic dystrophy

The condition exhibits relatively constant factors:

Muscular inactivity often precedes the onset.

Tenosynovitis of the long head of the biceps is found in almost every case at operation.

Frozen shoulder develops only in joints with severe degenerative change.

The frequent association of frozen shoulder with cardiovascular disease and painful restriction of hand motion strongly suggests an irritative focus which exerts its influence through the sympathetic nervous system.

Author's Theory

The shoulder is a closely fitted joint. The humeral head barely clears beneath the coraco-acromial arch. When one examines this motion at operation under local anesthesia, the humeral head and the tuberosities are closely approximated to the acromion and the very rigid sharp-edged coraco-acromial ligament. In order for the prominences of the tuberosities to proceed beneath the arch, the head is depressed inferiorly away from the arch by contraction of the biceps, whose long tendon passes over the head.

The rotator cuff acts similarly in depressing the head but also fixes it against the glenoid while the deltoid abducts the arm. When the cuff or the intra-articular biceps tendon are rendered ineffective by tears, degeneration, elongation, plus superimposed muscle weakness (result of inactivity), the upward thrust of the deltoid acts alone and at about 45° of abduction, the tuberosities impinge upon the coraco-acromial arch, and further gleno-

humeral movement is impossible. Scapular motions alone effect further raising of the arm by the shrugging mechanism.

The cuff and the biceps tendon are exposed constantly to trauma of being forced against the arch. This causes reactive engorgement, edema, round-cell infiltration and degeneration. The consequent thickening of these tissues acts as an additional mechanical barrier to passage of the head beneath the arch. Attempts to do so accentuate the traumatic inflammation and cause further swelling and greater interference with movement. A vicious cycle is instituted.

If early rest is started, swelling subsides, gliding surfaces are preserved, and a full range of motion is obtained. If constant trauma continues, the tissues degenerate further, and attempts at healing by granulation tissue result in production of fibrous adhesions and adherence of all tissues, including the biceps tendon, the cuff, the bursa, the capsule, the humeral head and the acromion The chronic inflammatory process tends to spread to the rest of the capsule and distally along the tendon sheath in the bicipital groove. Finally, almost all movement is lost.

By persisting at motion and continuing pressure of the arch against the thickened tissues on the superior aspect of the head, attritional changes, thinning and eventual obliteration take place. The intra-articular tendon disappears, and with it the deep tendon pain sensation is gone. A defect in the cuff results. With deep tendon pain gone, the patient persists more readily at attempted motion, which is gained eventually by a progressive wearing down of all subacromial tissues, including the bone. The tuberosity is smaller and displays evidence of friction and compression, e.g , sclerosis, cystic changes. The inferior capsular tissues are stretched, and the rotator muscles and the deltoid are strengthened to compensate for the absent long head of the biceps. The original strength of abduction is never regained Although a great recovery of functional range is obtained, some limitation of motion and weakness remains, the result of loss of depressor power

The above process may be inaugurated by any infectious process or trauma which thickens the soft tissues overlying the head.

Inactivity with the arm held in adduction quickly weakens the small scapular muscles and the biceps. The lax capsule at the inferior aspect of the joint contracts and prevents descent of the head. The degenerative and fibroblastic reaction replacing the elastic components in the capsular structure is a natural accompaniment of advancing age and explains the predominance in older individuals of frozen shoulder, contraction of metacarpophalangeal joints, the stiff knee after fractures in the elderly, etc.

The question of reflex sympathetic dystrophy needs further study. After an injury to the extremity, whether trivial or severe, there occasionally results a condition of severe, diffuse, poorly localized pain at one or several points such as about the shoulder, the elbow and the hand, associated with soft-tissue swelling, thinning of the skin, slight cyanosis of the part, coldness, hypersensitivity, limitation of motion, and increased sweating of the hand. Trauma may or may not be at a distance from the dystrophic area. When the hand and the shoulder are involved, it is known as the *shoulder-hand syndrome.* There appears to be a disturbance set up reflexly through the central nervous system which affects the neurocirculatory elements to that section of the body.

Treatment

In the early stages of the disease, bed rest, heat and sedatives are prescribed. If pain is severe, icebags are applied temporarily. The arm should hang in the dependent position, the weight of the arm providing traction. Hydrocortone may be injected into the shoulder and also given by mouth; often the pain is dramatically relieved within a few days The medication should be continued as long as relief of discomfort and improvement in the range of motion are obtained. Occasionally, the cervical sympathetic may be blocked, but the procedure rarely gives more than temporary relief.

Pendulum exercises are practiced as soon as they can be tolerated. Later, pulley exercises are added All motion should not exceed the range which causes only a minimum of discomfort. Forceful movement and manipulation are contraindicated, because fibrous ad-

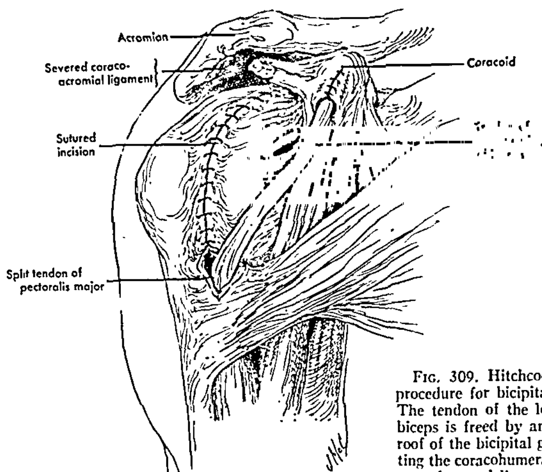


FIG. 309. Hitchcock and Bechtol procedure for bicipital tenosynovitis. The tendon of the long head of the biceps is freed by an incision in the roof of the bicipital groove and splitting the coracohumeral and the transverse humeral ligaments. The uppermost fibers of the tendon of the pectoralis major are severed for about 2 to 3 cm. The biceps tendon is severed at its attachment to the upper glenoid rim, removed from the joint and sutured to the coracoid and its attached tendons.

hesions and normal tissue are torn, further scar tissue is formed, and shoulder fixation becomes resistant to cure. The patient is advised that conscientious effort practiced daily over many months is the only effective method.

Surgical treatment is indicated (1) when pain is severe and persistent, (2) recovery of motion must be hastened, and (3) if a lesion of the cuff is suspected.

Technic.[4] A transacromial approach is used, and much of the acromion is discarded. The subacromial bursa is resected. Between the supraspinatus and the subscapularis portions of the cuff lies the coracohumeral ligament, which in this condition presents a cordlike thickening. The joint is entered by splitting this ligament longitudinally. The intra-articular biceps tendon is freed where it is adherent to the capsule and the head; its origin at the glenoid rim is cut; and the tendon is removed to the point where it enters the bicipital groove. The transverse humeral ligament is cut, the fascial roof is split, and the extra-articular tendon is elevated. If the tendon is to be fixed to the groove, all soft tissue is curetted from the groove, and the tendon is replaced and held by sutures run through adjacent drill holes. Otherwise, the tendon may be attached to the coracoid process.

Postoperatively, the arm is immobilized at the side for several weeks before graduated exercises are started. This procedure often dramatically effects recovery of motion and subsidence of pain.

[4] Hitchcock, H. H., and Bechtol, C. O.: Painful shoulder; observations of the role of the tendon of the long head of the biceps brachii in its causation, J. Bone & Joint Surg 30A 263, 1948.

DISLOCATION OF THE SHOULDER

These constitute about 50 per cent of all dislocations. The very redundant capsule, the

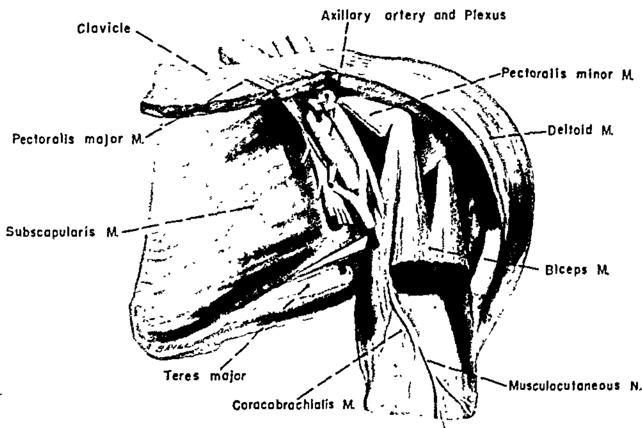


Fig. 310. Course and vulnerable position of brachial plexus and musculocutaneous nerve which may be injured at the time of the dislocation or by pressure of the head of the humerus left in a dislocated position for a long period of time. (DePalma, A. F.: Surgery of the Shoulder, p. 214, Philadelphia, Lippincott)

lax ligaments, the shallow glenoid cavity, all create an ideal setting for dislocation. The integrity of the surrounding muscles is important for stability and retention of the humeral head in the glenoid. Most dislocations occur after the age of 20, but are rare after 45 The most common types are the subcoracoid, the subglenoid and, very rarely, the subclavicular and the subspinous or posterior dislocation.

Mechanism

The displacement occurs by hyperabduction of the arm with the acromion acting as a fulcrum, and the head is levered out of the glenoid with tearing of the inferior capsule and stretching of the rotator cuff. Very rarely, a fall with the arm in forward flexion and marked internal rotation produces a posterior dislocation. A direct blow from behind may occasionally cause an anterior dislocation.

Pathology

DePalma observed the following at operation:

1. Stripping of the capsule from the inferior aspect of the humeral head
2. Disruption of the glenohumeral ligaments and stretching of the capsule enclosing the subscapular recesses
3. Labrum glenoidale sheared from the anterior glenoid rim
4. Tear or maceration of the labrum
5. Labrum not involved
6. Stripping of labrum, capsule and periosteum from the anterior surface of the neck of the scapula
7. Head lies extracapsular. Capsule is torn, or stripped from the head.
8. Head lies intracapsular, in the subscapular recess.
9. Tears of rotator cuff chiefly in subscapularis and supraspinatus. These varied from microscopic tears to gross tears.
10. Displacement of biceps tendon from the bicipital groove
11. Fracture of anterior rim of the glenoid
12. Severe stretching of the rotator mus-

cles, particularly the supraspinatus and subscapular muscles. This was a constant feature in all muscles explored.

COMPLICATIONS

The axillary nerve may be involved by direct pressure just proximal to its exit through the quadrilateral space. The resultant disability varies from slight weakness of the deltoid and hypesthesia over the upper outer aspect of the arm to complete paralysis of the muscle and anesthesia in the same area. This usually clears up in a matter of a few weeks to several months. Less commonly, the musculocutaneous nerve is involved and is manifest by paralysis of the biceps, the brachialis and

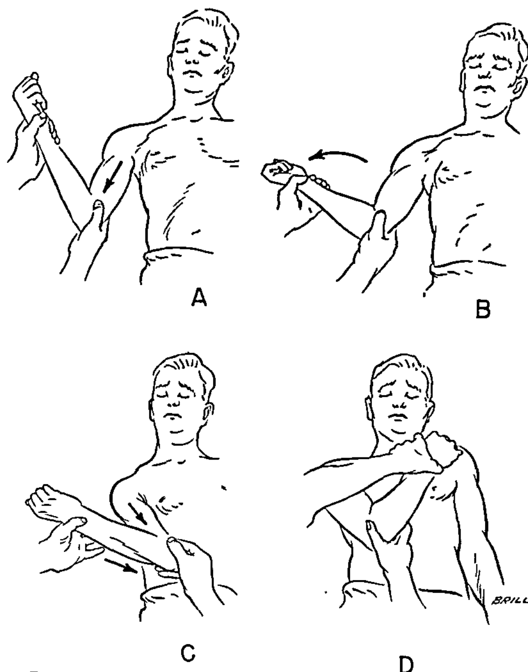


FIG. 311. Kocher's maneuvers to reduce dislocations of the scapulohumeral joint. (A) Preliminary stretching in the line of the long axis of the shaft of humerus in slight abduction. (B) The arm is externally rotated. (C) The elbow is moved forward to a point near the mid-line of the trunk. (D) Finally the arm is internally rotated so that hand falls over the opposite shoulder. (DePalma, A. F.: Surgery of the Shoulder, p. 218, Philadelphia, Lippincott)

the coracobrachialis and by reduced sensation along the outer border of the volar surface of the forearm. Rarely, the entire brachial plexus may be compromised, but fortunately this too is usually temporary.

Rarely, vascular injury occurs as manifest by a swollen, cyanotic extremity and an absent radial pulse The most commonly associated fracture is that of the greater tuberosity. Other types of fracture about the humeral head and the scapula rarely occur.

Clinical Picture

A history of a fall and immediate disability is often obtained. The patient holds the arm rigidly immobile in abduction and avoids any movement, which is excruciatingly painful. The distance from the acromion to the tip of the elbow is longer than on the uninvolved side. The normal rounded appearance of the shoulder is replaced by flattening below the acromion The head is palpable below the coracoid or the glenoid fossa When the rare type of posterior dislocation is present, the arm is held adducted at the side in internal rotation X-ray examination substantiates the diagnosis Roentgenographic studies are mandatory preliminary to instituting treatment to determine not only the type of dislocation but also the presence or the absence of fracture.

Treatment

The Kocher maneuver is used most commonly. This consists of flexing the abducted arm at the elbow, maintaining traction distally, slowly externally rotating the arm by using the forearm as a lever, bringing the externally rotated arm forward in front of the chest, and finally rotating the arm internally so that the hand is made to touch the opposite shoulder. A hand in the axilla manipulates the humeral head and aids its repositioning in the glenoid. Manipulation is done gently and slowly to prevent fracture, tearing of the cuff, and neurovascular injuries. The method of Hippocrates should be condemned, because it frequently causes damage to the axillary structures. It is performed by placing the foot as a lever in the axilla and exerting traction on the extremity.

Postreductive Treatment. Recurrence is common before the age of 30 but is rare after 45. However, a frozen shoulder is a common postdislocation sequela after 45. In young individuals, inasmuch as stiffness after prolonged immobilization rarely occurs, the extremity is splinted for several weeks to allow the injured tissues to heal. This is followed by gradually increasing motion and physiotherapy. The resumption of motion at first is permissible in all directions except abduction. In the older age group, because of the danger of stiffness, a collar-and-cuff sling is applied, and pendulum exercises are started immediately, with due care to avoid abduction. Early motion permits absorption of hemorrhage and exudate and results in early return of function.

Open Reduction of the Acute Dislocation

If closed reduction cannot be effected easily, a complicating factor, such as an interposed biceps tendon or capsule, which prevents reduction, should be suspected. The shoulder should be explored, preferably by the transacromial approach. Cuff tears are repaired. A displaced biceps tendon is removed and fixed to its groove or transplanted to the coracoid. A tear of the capsule at the inferior aspect of the head need not be repaired, because it heals readily.

Fracture of the Greater Tuberosity

Thirty per cent of dislocations are associated with fracture of the greater tuberosity which, upon healing, will impinge upon the acromion and obstruct abduction. If the fragment is in good apposition, it needs no special treatment. However, when it is displaced superiorly and posteriorly, reduction is accomplished by placing the arm in about 90° abduction (with care to avoid redislocation), 30° of forward flexion, and 60° of external rotation. If the tuberosity has not been fractured, the cuff adjacent to the tuberosity is usually torn This is manifest clinically by weakness of abduction of the arm, particularly against resistance, and by the so-called "drop arm sign." Tenderness proximal and superior to the greater tuberosity is excruciating. The actual extent of damage to the tuberosity and the cuff can be determined only by direct observation at operation. Open repair is more accurate and provides a more

satisfying functional result. The acromion should be resected to eliminate a potential obstruction to abduction.

Old Unreduced Dislocations

Failure of reduction of an acute dislocation is generally due to an interposed obstructing tissue such as a posteriorly displaced biceps tendon or massive avulsion of the cuff. If this condition persists or the original dislocation has been neglected, the humeral head becomes fixed by adhesions and scar contracture in its new situation beneath the subscapularis muscle. The adductor and the internal rotator muscles become fibrotic and shortened. A dense inelastic scar tissue fills the glenoid cavity and obliterates it. The musculotendinous cuff becomes contracted and shortened. The articular cartilage of the humeral head and the glenoid undergoes degenerative changes. The result is a shoulder which is painless or only slightly painful and has greatly restricted motion.

Treatment of Unreduced Dislocations. An attempt should be made to reduce the dislocation by manipulation when the condition has existed for no more than 3 months. The attempt should be done gently and cautiously to avoid fracture, cuff damage, and nerve and vascular injury. The reduction should be preceded by a period of traction followed by abduction and exernal rotation manipulation to stretch the subscapularis and the pectoralis major, the chief obstacles to reduction. Under general anesthesia, a gentle Kocher maneuver is performed while an assistant controls the humeral head and pushes it toward the glenoid. Reduction is associated with tearing of adhesions and hemorrhage resulting in marked shoulder stiffness Failure of reduction by the closed method is frequent. The alternative is to do an open reduction.

Open Reduction of an Old Irreducible Dislocation.[5] The incision skirts the margin of the acromion and descends along the deltopectoral interval. The deltoid muscle is reflected downward. The subscapularis tendon is cut, and the muscle is reflected medially, exposing the subscapular space in which the head lies surrounded by adhesions. The glenoid cavity is covered with scar tissue. The axillary nerve is isolated and protected. All fibrous tissue is removed, and the head is levered back into the glenoid cavity. Too much capsule should not be removed, as it deprives the head of its vascular supply and eventuates in avascular necrosis and a stiff painful shoulder. If the biceps tendon is displaced posteriorly, it is cut, and its proximal portion is inserted in a hole drilled in the humeral head and fixed to the distal segment as in the Nicola procedure. The coracohumeral ligaments may be cut from the coracoid and attached through a drill hole in the acromion to act as a suspensory ligament, preventing subsequent dislocation. Several Kirschner wires may be passed from the humeral head to the acromion to provide additional fixation; they are removed several weeks later. The capsule is closed, and the divided tendons are resutured. Postoperatively, the arm is immobilized in a cast in approximately 60° of abduction and midway between the sagittal and the frontal planes. After 4 to 6 weeks, intense physical therapy is started and includes graduated active exercises. If a painful, stiff shoulder ensues, arthrodesis in the functional position is indicated. In aged individuals with a painful unreduced dislocation or with severe degenerative arthritis, it is preferable to resect the humeral head and accept a flail shoulder joint.

[5] Cubbins, W. R, Callahan, J J, and Scuderi, C S: Irreducible shoulder dislocations, Surg, Gynec & Obst. 58 129, 1934.

Recurrent Dislocation of the Glenohumeral Joint

In young individuals, dislocations are prone to recur particularly if the initial episode is not followed by prolonged immobilization that enables the injured tissues to heal, and if muscle strengthening exercises are not done. Each subsequent dislocation occurs with greater ease and less pain, and reduction is less difficult until eventually reposition can be effected without anesthesia. This latter fact infers that marked stretching and laxity of the soft tissues have occurred.

The causes of recurrences and the reasons for failure of various operative procedures designed to stabilize the joint include: (1) detachment of the labrum from the anterior glenoid rim (Bankhart); (2) failure of heal-

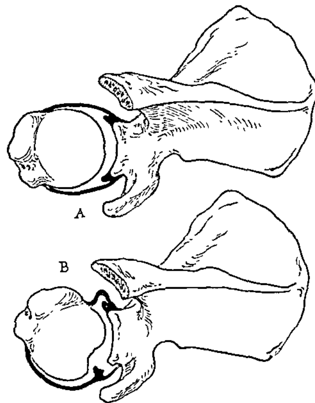


FIG. 312. Pathology of recurrent dislocation of the shoulder. (A) Normal appearance. The labrum attaches to the margin of and deepens the glenoid. (B) Pathologic appearance. A defect in the posterolateral aspect of the humeral head, avulsion of the labrum from the anterior lip of the glenoid, and stretching and separation of the capsule from the anterior neck of the glenoid. These factors contribute to but are not necessarily the primary causes of recurrent displacement. Not shown is the relaxed elongated subscapularis tendon which lies anterior to and fuses with the anterior capsule. Its relative lengthening permits excessive external rotation of the humeral head, a necessary prerequisite to dislocation.

ing of the torn capsule; (3) bony defect on the posterior aspect of the humeral head (Broca and Hartmann); and (4) laxity of the musculotendinous cuff. DePalma observed that the short rotators were stretched and the cuff was lax. The labrum was often detached from the anterior and the antero-inferior portions of the rim The labrum with the capsule and the periosteum was stripped from the anterior surface of the neck of the scapula. Normally, the anterior capsule is projected forward to attach beyond the neck, then is reflected back along the neck and attaches to the rim and the labrum. This forms the subscapular recess, which is stretched to accommodate the dislocated head. The thickenings within the capsule, the anterior and the inferior glenohumeral ligaments are stretched or torn, thereby removing the restraining barrier to dislocation. When the labrum is detached, the anterior rim is eroded and eburated. A

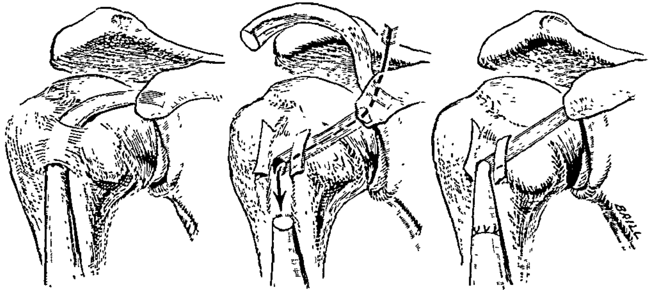


FIG. 313. The Nicola Operation The biceps tendon is converted into a suspensory ligament for the head of the humerus (DePalma, A. F.: Surgery of the Shoulder, p. 243, Philadelphia, Lippincott)

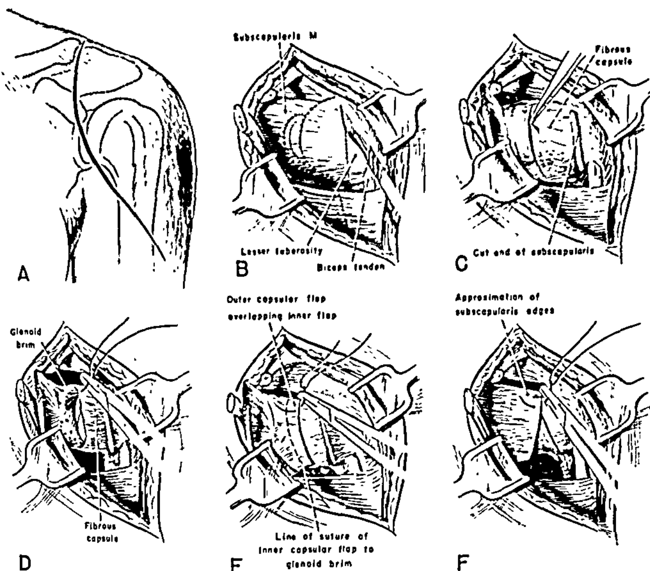


FIG. 314. Bankart Operation. (A) Deltopectoral incision (an S-shaped skin incision may be substituted). (B) Coracoid process has been sectioned and retracted medially with the coracobrachialis and short head of the biceps tendon. The subscapularis tendon is sectioned ½ inch from its bony insertion. (C) The subscapularis muscle is peeled off the fibrous capsule and retracted medially; the capsule is opened by a 2 inch vertical incision 1 centimeter from the glenoid margin. (D) Four drill holes are made in the glenoid brim, and the lateral capsular flap is sutured to the bony margin of the glenoid fossa. (E) Medial capsular flap is overlapped and sutured to the lateral flap. (F) Divided ends of the subscapularis tendon are approximated by interrupted silk or cotton sutures. (DePalma, A. F.: Surgery of the Shoulder, p. 240, Philadelphia, Lippincott)

posterolateral defect in the head is present in some but not all cases.

Normally, some degree of labral detachment is present after the third decade and is easily demonstrable in all subjects after the sixth decade. Yet recurrent dislocations are common in the early decades but rare after the fourth. Therefore, it is obvious that labral detachment and head defect are not causative factors. *The neuromuscular imbalance caused by severe stretching of the short rotators, particularly the subscapularis, is the main cause.* Capsular stretching is a secondary adaptive change. The changes described by other writers are normal degenerative wear and tear changes and may even be caused by trauma.

Surgical Treatment. The following are the procedures in common use:

NICOLA. The tendon of the long head of the biceps is fixed to a groove or drill-hole in the

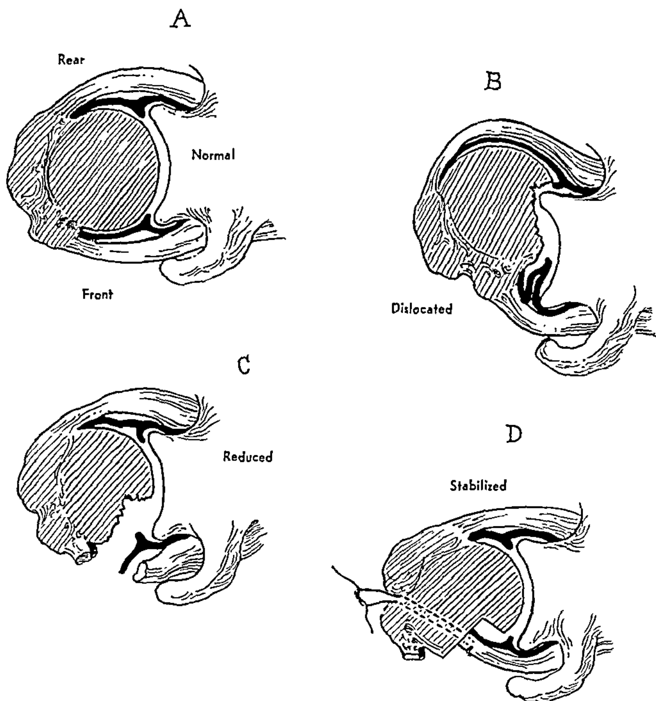


FIG. 315. Surgical correction for recurrent posterior dislocation of the shoulder. (A) Supero-inferior view of normal shoulder. (B) Posterior dislocation. The head is held in internal rotation by the engagement of the bony defect of the head against the posterior glenoidal rim (C) The reduced position. The bony defect is observed after cutting the tendon of the subscapularis and the capsule. (D) Final repair. The tendon and the capsule are sutured to the floor of the bony defect, thereby restricting internal rotation (McLaughlin, H. L.: Posterior dislocation of the shoulder, J. Bone & Joint Surg 34A:584)

head and the neck of the humerus However, the chronic pull on the site of attachment of the tendon leads to detachment of the labrum and may be the cause of failures.

BANKART. This is based on the premise that labral detachment is the causative lesion. The labrum is sutured back; or, in the event that the labrum is attenuated, the capsule

is sutured to the scarified glenoid margin.

PUTTI-PLATT. The subcapsularis tendon is severed, and the lateral segment is attached to the anterior capsule in front of the glenoid rim. The medial segment is lapped over and sutured to the lateral portion. This constitutes reefing of the tendon and in effect limits external rotation, a movement necessary to dislocation.

MAGNUSON. The insertion of the subscapularis is removed and reattached further laterally, thereby greatly restricting external rotation. The results are excellent.

POSTERIOR DISLOCATION OF THE SHOULDER[6]

This constitutes about 4 per cent of all types of dislocation of the shoulder. Clinically the diagnosis is made by a history of trauma to the adducted, internally rotated extremity, increased prominence of the coracoid, decreased prominence of the humeral head, marked limitation of abduction, absence of external rotation, a fixed internal rotation deformity, and by roentgenograms. The films are taken with the patient erect and facing the casette obliquely so that the central ray corresponds to the long axis of the scapular spine. The head of the humerus is revealed displaced posteriorly.

Treatment. Early, reduction is accomplished under general anesthesia by traction, external rotation and pushing the humeral head forward. Old unreduced dislocations require open reduction. The subscapularis is found to be stretched around the anterior rim of the glenoid. The glenoidal labrum is intact except for an abraded area in the posterior segment. The dislocated head is jammed tightly between the posterior margin of the acromion and the posterior rim of the glenoid. Its abnormal position is firmly fixed by the engagement of the posterior glenoid rim into a deep vertical defect in the anterior aspect of the humeral head, just medial to the lesser tuberosity. The subscapularis tendon is divided as close to its insertion as possible and implanted into the defect in the head. Failure to fill this defect allows the dislocation to recur with flexion and internal rotation. In some cases a posterior bone block is necessary in addition to the tendon transplantation.

SCAPULOCOSTAL SYNDROME[7]

The bursae and other soft tissues in the scapulothoracic interval are sensitive to abnormal pressure and friction which result from poor mechanical approximation of the scapula to the thoracic wall. Pain is referred to the cervical area, the shoulder, the chest and the hand.

CLINICAL PICTURE

The onset of pain is insidious and occurs in the superior or the posterior aspect of the shoulder girdle. The discomfort radiates to (1) neck and occiput, (2) upper triceps and deltoid insertion, (3) around the chest to the anterior aspect and (4) the medial forearm to the hands and the fingers, where numbness and tingling are complained of. The course is chronic and characterized by exacerbations and remissions.

ETIOLOGY

The cause is an alteration of the scapulothoracic relationship. The stimulus for the reflex originates in the extremely sensitive tissues beneath the scapula or in the suspensory muscles of the scapula. The condition is very common. The following types may be identified:

1. **Primary.** Poor posture causes poor fit of the retracted scapula. Characteristically, it is worse with fatigue toward the end of the day.
2. **Secondary.** The arm-trunk mechanism is not used for a prolonged period because of an associated condition, e.g., fracture.
3. **Static.** In the severely disabled, e.g., amputees whose deformities allow the scapula to sag or rotate.

DIAGNOSIS

There is diminished excursion of the scapula on the symptomatic side. The scapula may be retracted by having the patient place the hand of the involved side on the opposite shoulder. Then a trigger point may be located by pressing a finger against the chest wall

[6] McLaughlin, H. L.: Posterior dislocation of the shoulder, J. Bone & Joint Surg 34A 584, 1952.

[7] Russek, A S: Scapulo-costal syndrome, J.A.M.A. 150:25, 1952.

medial to the vertebral border of the scapula. The tender spot usually lies about the superior angle or base of the scapular spine. The suspected area is identified as the trigger point when pain is relieved by injection of local anesthetic into this site. On the other hand, local and reflex pain is intensified when distilled water or hypertonic saline is injected into the trigger zone.

Treatment

Local anesthesia breaks the reflex. A prolonged acting substance such as Intracaine in oil is used Multiple punctures may theoretically destroy inflamed distended bursae. Posture is corrected by exercises and supports. Secondary types require relief of the causative condition. Heat and massage are contraindicated, because the symptoms are made worse.

SNAPPING SCAPULA[8]

Any alteration in the smooth relationship of the scapulothoracic articulation causes audible and palpable sounds which are often painful and annoying to the patient.

Etiology

1. **Subscapular Bursae.** These cause a soft friction sound and the scapulocostal syndrome.

2. **Anterior Angulation of Superior Scapular Angle.** This is identified by an oblique roentgenographic view of the scapula and is confirmed at operation under local anesthesia. By elevating the medial border of the scapula, the snapping sensation and the pain are relieved.

3. **Tubercle of Luschka.** The superior angle may be abnormally enlarged by a small bony or fibrocartilaginous nodule situated on its anterior aspect. Usually this is identified only at operation.

4. **Scapular Osteochondromata.** Only those which develop on the anterior surface of the scapula give rise to snapping. Because they are compressed in the scapulothoracic interval during development, they present a characteristic mushroom-shaped appearance

5. **Omovertebral Bone.** An abnormal osseofibrous connection between the superior angle and the cervical spine represents the remains of an embryonic connection. This causes a grating sound and often winging of the scapula.

6. **Tumors and Deformities of Ribs.**

[8] Milch, H Partial scapulectomy for snapping scapula, J. Bone & Joint Surg 32A 561, 1950

Treatment

For anterior angulation of the scapula, the superior angle is resected. At operation, a tubercle of Luschka may be found and removed. Osteochondromata are removed and examined for malignant degeneration. The omovertebral bone is resected from its attachments at the cervical spinous processes and the superior scapular angle. When in doubt as the cause of snapping, the suspected site should be explored under local anesthesia by a muscle-splitting incision and the patient asked to move the part.

STERNOCLAVICULAR JOINT

The sternoclavicular joint is the pivot upon which the shoulder girdle moves on the trunk. It is a diarthrodial joint, permitting rotary movement of the clavicle. Internal rotation of the arm is associated with the maximum degree of rotation of the clavicle so that the bone protrudes anteriorly. A sharp, strong forward thrust of the shoulder girdle, as in the motion of throwing, is dependent upon the integrity of this joint. Elevation of the arm above 110° is associated with clavicular rotation. It is evident that restriction of motion at the sternoclavicular joint by surgery, trauma, or disease will greatly interfere with these functions.

Anatomy

The sternal facet at the upper angle of the manubrium faces posterolaterally. The cartilage-covered inner bulbous end of the clavicle articulates with the sternal facet and directly below with a facet on the first rib. A biconcave fibrocartilaginous meniscus is interposed between sternum and clavicle, forming two separate synovial-lined cavities. The meniscus is attached to the interclavicular ligament above and the sternochondral junction of the first rib below. A fibrous capsule encloses the joint and is strengthened anteriorly and posteriorly by the sternoclavicular ligaments. The capsule

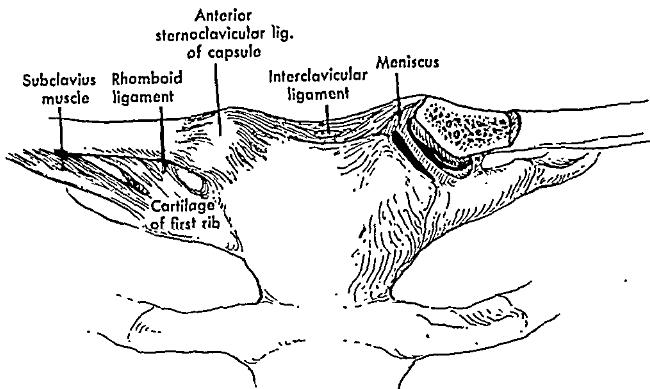


FIG. 316. The sternoclavicular joint. Note the capsule and the meniscus, both of which may become torn and interposed, preventing reduction. The tendon of the subclavius runs along the lower border of the muscle from which it may be separated and utilized as a sling to encircle the clavicle after reduction of a dislocation.

is especially thinned superiorly and inferiorly, the situation predisposing to dislocation. The rhomboid ligament fixes the inner end of the clavicle to the chondral portion of the first rib. It is often torn in sternoclavicular dislocations. The subclavius is a triangular muscle attached to the under aspect of the clavicle and inserting by a tendon to the costochondral junction of the first rib. Anteriorly, the sternoclavicular joint is covered over its medial aspect by the sternal head of the sternocleidomastoid muscle Posteriorly, it is in relationship to the sternohyoid muscle beneath which lie the trachea and the large vessels of the neck. These are compressed in posterior dislocations.

Dislocations of the Sternoclavicular Joint

The inner end of the clavicle most commonly is dislocated anteriorly and lies in front of the manubrium. The capsule may be torn and interposed so as to prevent reduction. Reduction, if possible, is effected by abducting the arm and pulling the shoulder girdle backward, at the same time manipulating the clavicle backward into the sternal facet. The patient is kept flat on his back with a sandbag placed over the clavicle. The arms are held at the side, and only minimal movement of the hands is permitted. It is extremely difficult to maintain this position for the 3 weeks necessary for capsule healing. Instability of the joint and redislocation are frequent occurrences. Surprisingly, many persistent dislocations permit rotation of the clavicle and excellent function. Occasionally, the displaced bone becomes firmly bound down with adhesions. Movement of the shoulder is painful and restricted, particularly internal rotation and abduction above 110°. Surgery is indicated for (1) pain, (2) restricted motion at the shoulder, (3) deformity and (4) as an emergency in posterior dislocations with compression of the great vessels. One may reduce the dislocation and provide stability by a fascial sling between clavicle and first rib, or by tenodesis, utilizing the subclavius tendon. If the dislocation is old and degeneration of the joint is probable, resection of the inner end of the

clavicle should be done. This is compatible with excellent function. The strength of abduction above the horizontal and of throwing is somewhat reduced

Bankart's Reconstruction[9]

Through a transverse incison, the dislocated clavicle is freed of adhesions. The interposed capsule usually lies behind the clavicle and is lifted upward, exposing the interior of the joint. If the meniscus is intact, it is left undisturbed; otherwise, it is removed. Capsule and periosteum are elevated as a flap from the front of the inner end of the clavicle and from the front of the sternum. The two flaps are turned down. By careful blunt dissection, the superior and the posterior aspects of these bones are stripped of soft tissues. A flat, broad spatula is placed behind the joint for protection. Two holes are drilled from before backward in the clavicle and also in the sternum near the articular margins. A tendon carrier is inserted in one hole in the clavicle, and one end of a fascia lata strip is pulled from behind forward. The other end of the strip is drawn similarly through the other hole in the clavicle. Then the two fascial ends are passed backward through the posterior capsule, then behind the sternal holes, through which they are again passed anteriorly. The clavicle is pushed backward, and the fascial strip is pulled up tight. The free ends may be tied, sewn together, or sutured to the soft tissues. The capsule and the periosteum are closed firmly.

Subclavius Tenodesis[10]

A transverse incision is made 1 inch below and parallel with the clavicle. Skin, superficial fascia and platysma are divided. The periosteum over the clavicle is elevated. The capsule is divided laterally and inferiorly, and the flap is turned medially. The pectoralis major is detached from the clavicle and the sternum and reflected downward, protecting the subclavius insertion. The subclavius tendon is dissected from its muscle fibers throughout its length, with special care to avoid damaging the subclavian vein. At the first costochondral junction, the tendon insertion is preserved. A hole is drilled in the clavicle anteriorly and again superiorly in line with the tendon insertion. The dislocation is reduced, and the tendon is brought through the anterior hole, emerges from the superior hole downward in front of the bone and is sewn to itself. The capsule is brought laterally and inferiorly and sewn over the front of the joint. The wound is closed in layers.

ACROMIOCLAVICULAR JOINT

The acromioclavicular joint is a hinge joint by which the clavicle articulates with the scapula. The flat articular surface of the outer end of the clavicle is directed outward, backward and downward to meet an acromial facet. A capsule and the acromioclavicular ligaments surround the articulation. Occasionally, a triangular meniscus is interposed between the facets. The acromioclavicular ligaments are weak and easily ruptured. However, stability of the joint is maintained by strong coracoclavicular ligaments, the conoid and the trapezoid ligaments, which firmly hold the clavicle down. The under surface of the joint is in relationship to the subacromial bursa and the musculotendinous cuff. Inflammation and spur formation, such as occur in degenerative arthritis of the acromioclavicular joint, traumatize and cause symptoms referable to the bursa and the cuff. Motion takes place at this joint chiefly with elevation of the arm above the horizontal plane, the movement being both rotatory and hingelike. When the joint is obliterated by surgery, injury, or disease, elevation of the arm above the horizontal is restricted.

Subluxation of the Acromioclavicular Joint

Direct trauma, such as an object striking the shoulder from above, or indirect trauma, such as a fall on the outstretched arm, may cause disruption of the acromioclavicular ligaments. The conoid and the trapezoid ligaments remain intact and prevent upward displacement of the clavicle However, a slight upward displacement is evident clinically. The

9 Bankart, A. S. B.: Recurrent sternoclavicular subluxation and dislocation, Brit. J. Surg. 26 320, 1938.

10 Burrows, H. J.: Tenodesis of subclavius in the treatment of recurrent dislocation of the sternoclavicular joint, J. Bone & Joint Surg 33B·240, 1951.

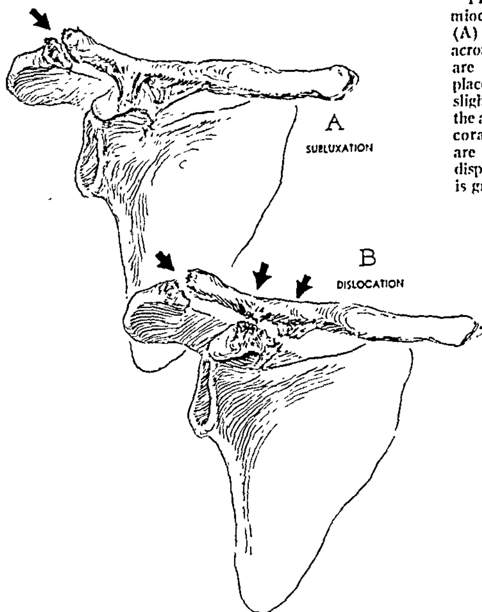


FIG. 317. Types of acromioclavicular separation. (A) Subluxation. Only the acromioclavicular ligaments are torn, and upward displacement of the clavicle is slight. (B) Dislocation. Both the acromioclavicular and the coracoclavicular ligaments are disrupted, and upward displacement of the clavicle is greater.

joint is swollen and tender, and movement at the shoulder is painful. An x-ray film usually fails to reveal anything abnormal. By taking an anteroposterior view of both shoulders while in the standing positions and both arms weighted with sandbags, the affected shoulder will be pulled downward, and the acromioclavicular interval will be widened as compared with the normal shoulder. As a rule, pain, swelling and tenderness subside in a few weeks, and normal function is restored. However, if the joint surfaces have been severely traumatized, degenerative changes supervene over the ensuing months. This is manifest clinically by symptoms of pain, stiffness, and restriction of arm abduction. Resection of the outer end of the clavicle relieves pain and restores full motion.

Dislocation of the Acromioclavicular Joint

A more intense trauma ruptures the coracoclavicular ligaments in addition to the acromioclavicular ligaments. The weight of the extremity causes the shoulder to droop downward, forward and medially. The clavicle is markedly displaced upward and backward, coming to lie posterior to the acromial facet. Clinically, the very obvious bony prominence above and behind the acromion is quite movable to manipulation. Initially, pain, tenderness and swelling occur but subside in a matter of a few weeks X-ray films show a separation of the coracoclavicular interval as compared with the opposite shoulder. The acromioclavicular interval is widened, and the superior margin of the clavicle lies well above

that of the acromial facet. The subsequent course of the untreated case is a shoulder with a good range of motion except hyperabduction. However, complaints of weakness, instability, fatigue and pain are common. Occasionally, the torn coracoclavicular ligaments are replaced by ossification which immobilizes the clavicle. The result is marked interference with movements of the shoulder, especially abduction and rotation. Correction of the dislocation and stabilization of the clavicle are essential.

Conservative Treatment

Reduction is easily effected by pressing the clavicle downward and forward while elevating the acromion by upward pressure on the elbow. A strapping is applied, encircling the clavicle above and the elbow below. Immobilization is continued for about 5 weeks. The strapping must be tightened every few days. However, it is extremely difficult to maintain the reduction, especially if soft-tissue interposition prevents apposition of the articular surfaces. In addition, the severity of the trauma often damages the articular cartilage and, when present, the meniscus also, leading to degeneration. For these reasons, open reduction is advisable.

Surgical Treatment

Recent tears of the coracoclavicular ligaments will heal if reduction is effected and maintained. Suture of the capsule alone is inadequate.

TECHNIC. The joint is exposed by an incision extending over the outer end of the clavicle to the edge of the acromion. The interposed capsular ligaments are elevated, and the joint surfaces are inspected. Fragments of cartilage and, if necessary, a damaged meniscus are removed. The clavicle is restored to its normal anatomic position and held by 2 threaded pins passed across the joint from acromion into the clavicle. The outer ends of the wires are cut off so as to lie beneath the skin and permit removal. The capsule is sutured. Postoperatively, a Velpeau bandage or a sling restrains the arm for about 4 weeks, after which restricted movement is permitted. At 8 weeks, the pins are removed, and full use of the extremity is allowed.

Old tears require some procedure which will replace the destroyed coracoclavicular ligaments and restore stability to the clavicle. This is especially necessary for athletes and those indulging in strenuous occupations.

TECHNIC. An incision is made over the outer third of the clavicle and over the joint to the outer edge of the acromion. The periosteum is elevated from the clavicle. Interposed capsule and fragments are removed from the joint. The anterior portion of the deltoid is severed from the clavicle and the acromial margin and reflected downward to expose the coracoid. Heavy braided silk is passed about the clavicle and, by a ligature carrier, is looped beneath the coracoid, exercising due care to avoid injury to the brachial plexus and the subclavian vessels. While the clavicle is pressed down in accurate reduction, the silk loop is pulled up tight and tied. Another braided silk loop may be added as a precaution. The capsule is repaired, the deltoid sutured back in place, and the wound closed. The silk acts as a foreign body about which fibrous tissue is laid down. The result in effect is the formation of strong stabilizing ligaments which permit full use of the extremity in about 2 months. A valuable substitute is to use half the width of the conjoined tendon of the short head of the biceps and the coracobrachialis. The tendon is severed longitudinally, left attached to the coracoid, looped about the clavicle, and sutured to itself.[11] Fascia lata, when used to replace the coracoacromial ligaments, will stretch and gives imperfect results. A transfixion screw passed from the clavicle to the coracoid has been used. However, it does not permit of clavicular movement and therefore restricts abduction and rotation of the arm. For the same reason arthrodesis of the acromioclavicular joint is contraindicated. If disruption of the joint indicates that degenerative arthritis is inevitable, it is advisable to resect the outer end of the clavicle distal to the point of attachment of the new coracoclavicular ligaments.

TUBERCULOSIS OF THE SHOULDER

Tuberculosis of the shoulder is rare and perhaps more frequent in adults than children. It seldom exists as a primary synovitis.

[11] Vargas, L.: Repair of complete acromioclavicular dislocation; utilizing the short head of the biceps. J. Bone & Joint Surg 24:772, 1942.

Rather, the lesion is found in the anatomic neck of the humerus and the glenoid of the scapula. The joint capsule is filled with masses of granulation tissue. In children the lesion may be metaphyseal and entirely extra-articular. Smaller discrete foci may become confluent and form a large fibrocaseous cavity which modifies the contour of the humeral head. A marked tendency to fibrous ankylosis and capsular contracture exists so that stiffness and marked restriction of motion rapidly appear. Longitudinal growth of the humerus occurs predominantly from the upper epiphyseal plate. Therefore, shortening results from tuberculous infections of the upper humerus in children. Atrophy of the deltoid and the periscapular muscles is marked and early.

Roentgenologic Findings

Early in the disease only osteoporosis is evident. The cortical margins are indistinct. The capsular shadow is expanded and denser than normal. The localized osteolytic lesions and narrowing of the articular space become apparent when the disease is well advanced.

Clinical Picture

Muscle weakness, a sensation of heaviness, and pain on motion are the earliest symptoms. Later, the pain becomes severe, and muscle spasm fixes the shoulder in adduction. The soft tissues become swollen, thickened and generally tender. Motion in all directions, particularly external rotation and abduction, is restricted. Marked muscle atrophy is present. The axillary lymph nodes are enlarged. Very rarely is a cold abscess found.

Treatment

Conservative treatment includes constitutional measures (nutritious diet, fresh air, bed rest, hygiene, vitamins), drugs (streptomycin, PAS, INAH) and immobilization by a shoulder spica cast. The shoulder is placed in the position of function in the event that ankylosis should supervene; 90° of abduction, 60° of external rotation and 30° of forward flexion is best. In adults it may be advisable to limit abduction to 60° or 70° to allow the arm to be brought down to the side.

A sound bony ankylosis is the only means of permanent cure. When general constitutional improvement permits, the abnormal tissues are resected, a combined intra-articular and extra-articular arthrodesis is done, a shoulder spica is applied, and immobilization is continued until bony bridging of the joint is demonstrable by roentgenograms. The operative procedures are described in the section on Poliomyelitis.

Under rare conditions, it may be desirable to preserve shoulder motion. Resection of the upper end of the humerus leaves a movable but weakened shoulder. The risk of future exacerbation is ever present.

22

The Elbow

SURGICAL ANATOMY

Bones

The lower extremity of the humerus and the upper extremities of the radius and the ulna form an articulation which is a diarthrodial ginglymus or hinge joint. The shaft of the humerus, which is triangular in cross section at its middle, becomes flattened anteroposteriorly and broadened transversely as its distal end is approached. Two thick condyles, medial and lateral, composed of cancellous bone, form the distal extremity of the humerus The lateral condyle is surmounted by the *capitellum*, a dome-shaped prominence which articulates with the shallow concavity on the radial head. Just above the capitellum on the anterior surface of the humerus is a shallow fossa which accommodates the edge of the radial head on full flexion of the elbow. Hypertrophic bone formation at this site following a supracondylar fracture will obstruct flexion movement. The posterior aspect of the capitellum, since it is not an articular surface, lacks a cartilage covering.

The *trochlea* is the spool-shaped process surmounting the medial condyle which articulates with a notch in the upper extremity of the ulna. The medial rim of the trochlea is prominent, extending a little distalward. Articular cartilage completely envelops the trochlear surface as far back as the olecranon fossa. Just above the trochlea the humerus is thinned out to form an anterior, or coronoid, fossa and a posterior, or olecranon, fossa. These accommodate the coronoid process of the ulna in flexion and the tip of the olecranon process in extension. Hyperostotic formations in these fossae or about the ulnar prominences will limit elbow motion in a corresponding direction.

Immediately above the condyles on the medial and the lateral aspects of the humerus are the *epicondyles*, bony prominences for tendinous attachment. The common tendon of origin of the flexor-pronator muscle group originates mainly from the very prominent medial epicondyle and from the medial supracondylar ridge which extends for a short distance above the epicondyle. Similarly, the extensor-supinator group arises from the smaller lateral epicondyle and the lateral supracondylar ridge. Both epicondyles lie outside the synovial cavity of the elbow. The collateral ligament on each side of the joint attaches to the respective epicondyle. When an epicondyle is avulsed from the shaft, the capsule and the collateral ligament are torn, exposing the interior of the joint into which the fragment may become displaced.

The condyles and their articular processes tilt forward, forming an angle of approximately 40° with the axis of shaft. In reducing supracondylar fractures in children, the ossification center for the trochlea must be angulated forward before reduction is considered to be adequate. Otherwise, permanent limitation of flexion will ensue. Conversely, excessive forward tilting of the epiphysis will limit extension.

The transverse axis of the lower humeral epiphyses lies obliquely so that the lower articular surface faces slightly outward. Therefore, when the elbow is fully extended, the forearm forms an obtuse angle with the arm of about 160°. In examining for this *carrying angle*, the arms are held adducted to the side, the forearms are supinated so that the palms of the hands face forward, and the deviation of the forearm away from the side is noted. Improper reduction of a fracture, or asymmetric epiphyseal growth following trauma or

infection, will produce a deformity in which the forearm deviates further outward (*cubitus valgus*), or further inward even to the point of reversal of the carrying angle (*cubitus varus*).

The expanded upper extremity of the ulna is formed anteriorly by the *coronoid process*, to which attaches the brachialis, and posteriorly by the *olecranon process*, the site of attachment for the triceps. Between the two is a deep semilunar notch, the *greater sigmoid fossa*, or incisura semilunaris, which articulates with the trochlea. Overlying the tip of the olecranon and firmly attached to the periosteum is the *olecranon bursa*.

Immediately distal to the greater sigmoid notch and situated on the lateral aspect of the ulna is a shallow groove, the *lesser sigmoid fossa* or incisura radialis. This cartilaginous covered surface receives and articulates with the periphery of the radial head.

The upper extremity of the radius is the disk-shaped *radial head* which is covered with cartilage and lies completely within the synovial cavity. Its proximal surface is concave for articulation with the capitellum. About 3 cm. beyond the lower margin of the radial head, the *bicipital tuberosity*, the site of insertion of the biceps, projects medially and anteriorly.

Capsular Ligaments and Synovium

The elbow joint is enveloped in a fibrous capsule which attaches proximally to the humerus just above the olecranon and the coronoid fossae, distally to the ulna just beyond the greater sigmoid notch, and to the neck of the radius and the lesser sigmoid notch. The capsule is thin, pliable and redundant anteriorly and posteriorly to permit free flexion and extension. Capsular thickenings, the collateral ligaments, on the inner and the outer aspects of the capsule, are tense and provide stability by preventing mediolateral motion.

The *medial (internal) collateral ligament* extends from the lower edge of the medial epicondyle and fans out to attach to the margin of the greater sigmoid fossa.

The *lateral (external) collateral ligament* attaches to the lower edge of the lateral epicondyle and passes distally to blend with the annular ligament.

The *annular (orbicular) ligament* is composed of transversely disposed fibers which encircles the radial head and attaches distally to the radial neck and medially to the anterior and the posterior margins of the lesser sigmoid fossa of the ulna.

Synovial membrane lines the inner surface of the capsule. Synovial sacs are formed proximally where it is reflected onto the humerus above the olecranon and the coronoid fossae and extends to the articular margin. Distally, the membrane forms a recess where it is reflected at the neck of the radius. In this latter situation, the radial head and a portion of the neck lie entirely intrasynovial, explaining the ease with which fracture fragments of the radial head are displaced.

Superficial Veins and Sensory Nerves

Lying anteriorly superficial to the deep fascia are large veins which are commonly used for venipuncture. These are the *cephalic vein*, lying laterally, and the *basilic vein*, lying medially which, with their communicating veins from the forearm, form the letter M. Medial to the cephalic vein lies the *lateral antibrachial cutaneous nerve*, and medial to the basilic vein lies the *median antibrachial cutaneous nerve*.

The Deep Fascia

A dense, tough, inelastic membrane lies beneath the veins and covers the muscles anteriorly and posteriorly. Medial to the biceps tendon it is reinforced by fibers arising from the biceps tendon, the *lacertus fibrosus*. This tough, unyielding fascia is of importance in supracondylar fractures when it retains extravasated blood under tension and compresses the neurovascular structures over the lower edge of the upper humerus fragment. Over the posterior aspect of the elbow the fascia covers the triceps expansion and with the latter attaches to the periosteum over the olecranon. Loose areolar tissue which covers the triceps fascia is often utilized for surrounding tendon grafts. From the medial and the lateral aspects of the deep fascia, the medial and the lateral intermuscular septum

passes inward to separate the anterior from the posterior arm muscle groups and attaches to the humerus.

Muscles

Posteriorly, the *triceps*, which attaches through its aponeurosis to the olecranon, effects extension of the elbow joint. Anteriorly, the *biceps*, which inserts into the bicipital tuberosity of the radius, is responsible for flexion and supination; and the *brachialis*, which inserts into the coronoid process of the ulna, assists in flexion.

Medial to the biceps and the brachialis, the *flexor-pronator group* of forearm muscles (pronator teres, flexor carpi radialis, palmaris longus, flexor carpi ulnaris, flexor digitorum sublimis) take origin by a conjoined tendon from the medial epicondyle of the humerus. Because these muscles assist in flexion of the elbow, this function can be utilized in paralysis of the biceps and the brachialis. By transferring the tendon of origin to a more proximal site on the humerus, flexor power is reinforced (Steindler procedure).

Lateral to the tendons of the biceps and the brachialis lies the brachioradialis, originating from the lateral supracondylar ridge of the humerus; and lateral to this is the extensor group of muscles of the wrist and the hand (extensor carpi radialis longus and brevis, extensor digitorum communis, extensor indicis proprius, extensor carpi ulnaris).

The Antecubital Space

The medial and the lateral forearm muscle groups approach each other and form a V-shaped interval (between the pronator teres medially and the brachioradialis laterally) in which lie important vessels and nerves. Medial to the biceps tendon is a neurovascular bundle consisting of the *brachial artery and veins* and

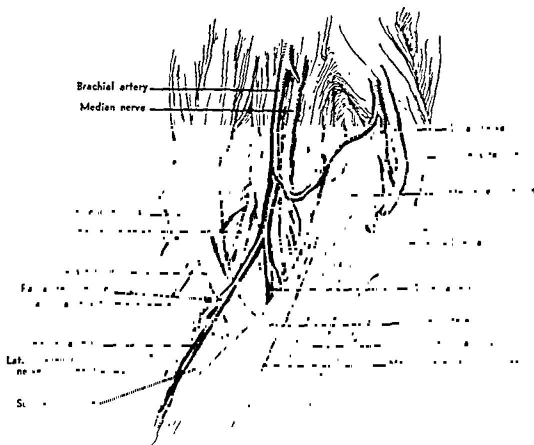


Fig. 318. The cubital fossa viewed from anteromedial aspect. Note particularly the profuse arterial anastomoses, the ulnar nerve lying superficially behind the medial epicondyle, the radial nerve running under cover of the brachioradialis, and the median nerve which disappears from view by exiting beneath the pronator teres

more medially the *median nerve*. The brachial artery divides into radial and the ulnar branches. The *radial artery* passes laterally and distally beneath the brachioradialis muscle. The *ulnar artery* passes medially and distally beneath the pronator teres.

The *median nerve* courses deeply into the antecubital space, passes behind the pronator teres and continues distally upon the surface of the flexor digitorum profundus. An injury of the nerve at the elbow will cause paralysis of the flexor digitorum sublimis, the radial half of the profundus, and the opponens of the thumb. An injury more distally will not affect the long flexors of the fingers.

The *radial nerve* runs in the interval between the brachioradialis and the brachialis. About 3 to 4 cm. above the joint, the radial nerve divides into (1) the *posterior interosseous nerve*, which runs distally to a point ½ inch below the radial head where it winds about the shaft of the radius, between the superficial and deep bellies of the supinator muscle, to reach the dorsal aspect of the forearm. (The nerve is easily injured by ill-advised surgical exposures about the radial neck, resulting in paralysis of the long extensors to the fingers and the thumb); (2) a *sensory branch* which courses distally beneath the brachioradialis muscle alongside the radial artery to reach the back of the hand and the thumb. Before dividing into its two main branches, the radial nerve supplies muscular branches to the brachioradialis and the radial wrist extensors. Therefore, the functions of these muscles often are unaffected.

The *ulnar nerve* runs posteriorly in a shallow groove behind the medial epicondyle of the humerus, then enters the front of the forearm between the heads of the flexor carpi ulnaris. The nerve is injured by irregularities in the groove or by stretching over a bony prominence or a cubitus valgus deformity as a result of a growth disturbance or malunited fracture. Symptoms of traumatic ulnar neuritis require dissecting the nerve from its groove and transplanting it to the anterior aspect of the elbow. The nerve should be freed upward to the lower third of the arm where it passes through the medial intermuscular septum. This septum must be resected. Otherwise, the nerve will be angulated as it is brought forward

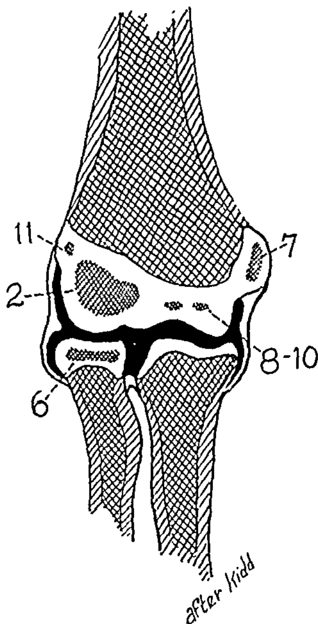


Fig. 319. The epiphyses about the elbow joint. The year of appearance of the ossification centers is indicated.

Bursae

Two bursae lie in relation to the triceps insertion. One lies between the triceps tendon and the upper surface of the olecranon. The other, the larger and more important one, lies between the skin and the dorsal surface of the olecranon. It is often inflamed and swollen as a result of trauma, infection and gout. A bursa lies between the neck of the radius and the biceps tendon as the latter approaches the bicipital tuberosity. Other bursae have been described about the lateral collateral ligament.

Epiphyses

In children the articulating processes at the ends of the long bones are composed mainly of cartilage. The junction of these cartilaginous masses with the bone of the shaft is a point of weakened resistance through which epiphyseal separations occur. Displacement of epiphyses can be recognized by identifying the individual ossification centers and noting their relationship to the parent bone. Therefore, it is important to know the time of appearance for these centers, their configuration, and the age at which they unite with the main bone. The elbow is an excellent site for determining bone age and various epiphyseal disturbances.

Ossification Centers of the Elbow

Center	Appears	Unites with Shaft
Capitellum and outer half of trochlea	2 yrs.	16 yrs
Trochlea, inner half	10 yrs.	16 yrs
Medial epicondyle	5 yrs	18 yrs
Lateral epicondyle	12 yrs	16 yrs
Radial head	6 yrs	18 yrs
Olecranon	10 yrs	16 yrs.

These figures are averages. Because considerable variation may occur, roentgenograms of the opposite elbow should be taken routinely for comparison This will avoid errors such as mistaking a displaced medial epicondyle for the trochlea.

Elbow Motion

The elbow is a diarthrodial ginglymus or hinge joint. Flexion and extension, or hinge motion, takes place between the lower end of the humerus and the upper ends of the radius and the ulna. In measuring the ranges of flexion and extension, full extension with the forearm lying in the axis of the arm is designated as 180°. The degree of flexion is measured by the angle which the forearm *forms with the arm as it approaches the latter*, the normal being 30°. In describing elbow motion, it is advisable to designate the arc through which the forearm passes from the extreme of extension to the extreme of flexion This is important in determining the usefulness of this range of motion. For example, 80° of motion is functional when the arc of motion is from 60° to 140°, whereas an arc from 100° to 180° would limit the patient's ability to eat or comb his hair with this hand.

Rotary motion (supination and pronation) takes place at the proximal radio-ulnar joint and at the distal radio-ulnar joint. At the elbow the edge of the disk-shaped radial head glides in the lesser sigmoid notch, pivoting about the prominence of the capitellum. During pronation, the distal end of the radius swings dorsally and comes to lie medial to the head of the ulna. Disruption of either the upper or the lower radio-ulnar joints or a disturbed capitellum-radial head relationship will limit rotary motion.

In examining for rotary motion of the forearm, the arm must be held adducted to the side, and the elbow is flexed to 90°. Otherwise, movements of the arm will contribute to positioning of the forearm, and accurate determinations will be impossible. The starting neutral position is assumed by placing the palm of the hand with the abducted thumb pointed toward the ceiling. From this zero position, the arcs of pronation and supination are measured. The usual normal pronation range is 90°, and the supination range is 90°, but variations are not uncommon. To determine whether or not limitation of motion is present, a comparison with the uninvolved extremity should be routine.

Bony Landmarks

Both epicondyles and the tip of the olecranon process form useful palpable bony prominences. With the elbow flexed to a right angle and examined from behind, an isoceles triangle is formed by lines drawn between the epicondyles and from each epicondyle to the olecranon tip. When the elbow is fully extended, the 3 bony points lie along a straight transverse line. Viewed from the lateral aspect, these points lie in the axis of the humeral shaft. These relationships are altered when a *fracture or a dislocation is present.*

On the lateral aspect of the elbow, the radial head is palpable and is felt to rotate with supination and pronation of the forearm. The radial head, the lateral epicondyle and the tip of the olecranon form a triangle over the posterolateral aspect of the joint. This interval is occupied by the anconeus muscle beneath which is the capsule of the

joint. When the joint is distended with fluid, the anconeus triangle bulges outward. Aspiration is conveniently accomplished at this site.

TENNIS ELBOW
(Epicondylitis; radiohumeral bursitis)[1-5]

The syndrome of chronic disabling pain in the elbow, particularly about the radiohumeral articulation, is designated "tennis elbow" rather than epicondylitis or radiohumeral bursitis in view of lack of specificity regarding its origin.

Etiology

The actual cause is unknown. It is common in individuals whose occupations require frequent rotatory motion of the forearm, e.g., tennis players, pipe-fitters, carpenters, etc. Frequently, it is associated with tendinitis of the shoulder, fibrositis of the back, and other collagenous degenerative conditions occurring in young and middle-aged adults.

Clinical Picture

The onset is gradual. An ache appears over the outer aspect of the elbow and is referred into the forearm. It is persistent and intensified by grasping or twisting motions. Grasping requires setting of the extensor carpi radialis brevis and longus; rotatory or twisting motions of the forearm toward supination require active contraction of the supinator longus (as well as the brevis). These muscles originate from the lateral epicondyle and epicondylar ridge and a few fibers from the anterior capsule of the elbow joint. A well-localized point of tenderness exists at one of the following sites: (1) epicondylar ridge, (2) lateral epicondyle, (3) lower edge of capitellum anteriorly, (4) laterally over the radiohumeral interval, and (5) one area in the circumference of the radial head during rotation of the forearm; most commonly is over the anterior aspect in the position of full supination. Swelling is rarely present, and the range of motion is normal. The patient complains of weakness of grasp and dropping of objects, particularly with the forearm pronated. A *clinical test* consists of reproducing the pain by completely extending the elbow, pronating the forearm and forcibly flexing the wrist. Active attempts to dorsiflex the wrist and supinate the forearm against resistance will likewise intensify the discomfort.

The condition infrequently involves the medial epicondyle. In this case, the pain and the tenderness exist about the medial epicondyle and the common flexor pronator tendon of origin. Activities which impose tension on this tendon accentuate the pain. The discomfort is intensified by strong grasping, active flexion of the wrist and pronation of the forearm against resistance.

Roentgenologic Findings

The roentgenograms are usually negative. Occasionally, a small flake of bone anterior to the epicondyle suggests an avulsion; or the surface of the epicondyle may be roughened as an indication of periostitis.

Pathology

The actual pathology is unknown. The majority of opinions indicate that the condition is caused by a partial tearing of the tendon fibers from their attachments to the epicondyle and the epicondylar ridge; the constant muscle contractions prevent healing, creating a traumatic periostitis. Therefore, treatment is directed toward (1) complete severance of tendon from its attachment, and (2) firm fixation of tendon origin.

Bosworth demonstrated that the annular ligament undergoes hyaline degeneration and may be the source of pain. He reports cures by resection of the ligament, but in addition he severed or resutured the common tendon.

Other reported pathologic conditions include: (1) arthritis of the radiohumeral joint, (2) radiohumeral bursitis, (3) traumatic synovitis of the radiohumeral joint through forced extension and supination, and (4) periostitis or osteitis of the epicondyle.

[1] Bosworth, D M. The role of the orbicular ligament in tennis elbow, J Bone & Joint Surg. 37A: 527, 1955

[2] Spencer, G E, Jr, and Herndon, C H. Surgical treatment of epicondylitis, J Bone & Joint Surg. 35A:421-424, 1953.

[3] Smith, C. H, and Kunz, H G: Butazolidin® in rheumatoid disorder, J. M Soc. New Jersey 49.306, 1952.

[4] Cyriax, J. H.: The pathology and treatment of tennis elbow, J. Bone & Joint Surg. 18:921, 1936.

[5] Mills, G P: The treatment of tennis elbow, Brit. M. J. 1:12, 1928

Treatment

Conservative. *This is effective in most* cases, but recurrences are common.

1. Rest. Complete rest by a posterior molded cast or splint, maintaining relaxation of the extensors by flexion at the elbow, supination, and extension at the wrist. The fingers may be left free for movement. The cast should be removed daily for gentle exercises to avoid elbow stiffness.

2. Heat. Moist compresses or short-wave diathermy

3. Procaine. Multiple punctures are made in the tender area and are repeated at intervals of 5 days to a week.

4. X-ray Therapy. Three treatments of 200 r in air to each of three fields—anterior, posterior, and lateral, one field being treated every other day. (Modality 220 KV, 0.5 mm. copper, 1.0 mm. aluminum filter with a half value layer of 1.2 mm. copper.)

5. Manipulation. The principle is to convert the partial tear of the conjoined tendon into a complete tear, thereby detaching the tendon from the chronically inflamed periosteum.

Technic. With the elbow flexed and the forearm supinated, the epicondyle is massaged for 10 minutes. Then the elbow is fully extended, and the forearm forcibly adducted to create a cubitus varus position. The treatment is repeated at intervals of 2 or 3 days; about 4 treatments are sufficient to provide relief.

Technic. While the fingers and the wrist are held fully flexed and the forearm pronated, the elbow is forced into full extension while firm pressure is applied with the thumb over the tender epicondyle.

6. Miscellaneous Ultrasonic therapy has produced equivocal results Butazolidin (phenylbutazone) produces excellent results in reported cases but is not recommended in view of its potential toxicity. Local injection of hydrocortone gives results no different than needling under local anesthesia.

Surgical. This usually gives immediate and lasting relief of symptoms. It is indicated when conservative treatment has failed

Technic. A tourniquet is applied. A curved linear longitudinal incision 1½ inches long is made just posterior to the lateral epicondyle and along the head of the radius. The deep fascial covering over the conjoined tendon is divided transversely. The intermuscular septum is also divided. The conjoined tendon is severed at the epicondyle and the epicondylar ridge, and detachment of the remaining fibers is completed by subperiosteal elevation, including their connection with the anterior joint capsule. The tendon and the extensor muscles are allowed to displace distally. A lateral incision into the joint anterior to the collateral ligament exposes the capitellum and the radial head. A portion of the annular ligament is resected. Only skin and subcutaneous tissue are closed. An elastic compression bandage is applied, and immediate motion of the joint is permitted. The removal of the annular ligament is optional and does not affect stability of the radial head. Without this step, results are satisfactory.

Technic. Firm fixation of the conjoined tendon is done to avoid the theoretical weakening of the extensor muscles resulting from reducing the distance from origin to insertion. The conjoined tendon is elevated at the epicondyle, and the cortical bone is curetted to expose cancellous bone. Crisscross sutures of heavy black silk are placed through the tendon, the end of the tendon is freshened, and the latter is approximated to the cancellous bone as the suture is drawn through drill holes. Postoperative immobilization with the elbow flexed, the forearm supinated, and the wrist dorsiflexed is maintained for 6 weeks.

ELBOW SPRAIN

Forced movement of the joint beyond full extension or forced abduction of adduction of the forearm tears the capsular apparatus and its contained reinforcing ligaments from its attachment to the humerus above the ulna and the radial head below.

Sprain of the Radial Collateral Ligament

This ligament extends from the lateral epicondyle to the side of the orbicular ligament. The tenderness is generally below the lateral epicondyle, and pain is accentuated by forcing the elbow into varus Treatment consists of immobilizing the elbow in flexion.

Sprain of the Ulnar Collateral Ligament

This ligament extends from the medial epi-

condyle to the ulna. Tenderness is below the epicondyle, and pain is accentuated by forcing the elbow into valgus. Treatment is by immobilization in flexion. More commonly, the ligament attachment is strong, and instead the medial epicondyle is avulsed and displaced downward. Healing frequently is fibrous but clinically satisfactory.

Sprain of the Anterior Capsule

Violent hyperextension of the elbow short of actual posterior dislocation causes pain and tenderness over the attachment of the capsule to the lower anterior aspect of the humerus. Pain is reproduced by attempting to extend the joint. Treatment consists of immobilization in flexion for a few days.

PULLED ELBOW
(Goyrand's Injury; Malgaigne's Luxation)[6]

Subluxation of the radial head distally to the annular ligament so that the latter becomes interposed between the radial head and the capitellum can occur by forcible traction distally on the radius. The displacement is possible only before the radial head has fully developed. Before the age of 6, the width of the radial head is flush with the shaft and offers no obstruction to slipping upward of the annular ligament. The injury is most common at the age of 2; typically, the parent forcibly pulls on the forearm in lifting the child to a higher level such as a curb or a step. Occasionally, a palpable or audible snap is perceived, the child cries out, and immediately the arm remains limp at the side with the forearm in neutral position or partial pronation and the elbow about 20° to 30° short of full extension. On examination, tenderness exists over the radial head, and one experiences a rubbery resistance to passive supination and extension of the elbow. Roentgenograms are negative. Reduction is performed without anesthesia. The radial head is forced anteriorly by thumb pressure as the forearm is slowly supinated and extended. Sudden release of resistance accompanied by a definite click signifies replacement. If this manipulation fails, one may try flexing the elbow and at the same time pronating and supinating the forearm. A sling is worn for 1 week. Recurrences are common and are similarly treated. Occurrence after 8 years of age is highly improbable.

[6] Magill, H. K., and Aitken, A. P.: Pulled elbow, Surg., Gynec. & Obst. 98:753, 1954.

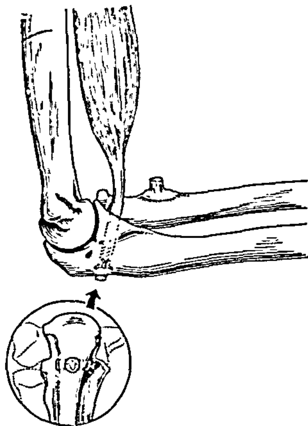


Fig. 320. Transplantation of biceps tendon. Reichenheim's technic modified by passing tendon through drill hole in coronoid to the subcutaneous border of the ulna, where it is sutured. (King, T.: Recurrent dislocation of the elbow, J. Bone & Joint Surg. 35B:50-53)

RECURRENT DISLOCATION OF THE ELBOW

The basic causes of this rare disability include an initial *severe injury* which effects a tear in the thin anterior capsule, torn collateral ligaments and a fracture of the coronoid process of the ulna; a *shallow semilunar notch* in the olecranon; and an *insufficient trochlea,* usually a result of fracture. The most commonly associated pathology is a fracture of the coronoid with upward displacement of the fragment, causing a shallow semilunar notch in which the trochlea cannot engage.

In the intervals between dislocations, the elbow feels weak and insecure. On examination the elbow can often be hyperextended. Abnormal medial and lateral mobility attests to inadequate collateral ligaments.

X-ray examination may reveal a shallow semilunar notch in the ulna, with or without a displaced coronoid fragment; or an inadequate trochlea with evidence of an old fracture or other causative condition such as osteochondritis dessicans.

Treatment

Surgical correction aims at restriction of hyperextension and, if possible, providing an anterior block to extension. The biceps tendon is removed from its insertion and drawn through a drill-hole made anteroposteriorly in the olecranon. Relative shortening of the biceps greatly limits elbow extension, but this range of motion is restored in time. Active supination is not affected.[7]

OLD UNREDUCED DISLOCATION OF THE ELBOW

An acute dislocation of the elbow, if neglected or following failure of reduction, will quickly develop intra-articular and periarticular adhesions, contracture of periarticular ligaments, and shortening of the triceps; after 2 weeks, closed reduction is virtually impossible. If closed reduction cannot be effected within the first 24 to 48 hours, open reduction is advisable, because forceful manipulative efforts will tear the soft, edematous periarticular soft tissues, including neurovascular structures, with additional hemorrhage and inevitable permanent stiffening of the elbow. Such ill-advised trauma is often followed by myositis ossificans. The earlier the operative intervention the better the results as regards preservation of motion. After reduction by either the closed or the open method, early exercise will prevent adhesions between apposed synovial surfaces. Such motion should be active and gentle, with surprisingly good restoration of the range of motion taking place slowly over a number of months. Forceful, passive motion, which attempts to overcome a resistance to flexion or extension, often causes worsening of joint stiffening.

The only contraindication to early surgical correction is the presence of myositis ossificans. Operative trauma encourages the spread of heterotopic ossification, the periarticular structures being infiltrated by the process, and a hopelessly stiffened joint is produced. The circumstances easily suggest to the patient that the surgeon has been at fault. If on the preoperative roentgenogram one finds the cloudy shadow of recent ossification in the soft tissues, it is advisable to defer surgery until the shadow becomes dense, well delineated and stationary. It may require many weeks or months, but then one can proceed with considerably less hazard. If the new bone formation constitutes an obstacle to elbow movement, it should be removed.

Technic

A longitudinal incision is made over the posteromedial aspect of the arm in the lower third. Just above the olecranon the incision is carried transversely to the lateral humeral condyle and then distally over the head of the radius into the forearm. The ulnar nerve is dissected from its groove behind the medial epicondyle and freed upward and downward and retracted out of harm's way. The aponeurosis of the triceps is dissected out as a tongue-shaped flap with the base attached to the olecranon distally. The triceps muscle is split in the mid-line and elevated, exposing the posterior joint capsule. Subperiosteal elevation of soft tissue is carried medially and laterally to the anterior surface of the humerus until all muscular attachments are freed. Next, the capsule is split posteriorly in the mid-line and detached completely from around the humerus. At this stage, one encounters much scar tissue and callus formation about the olecranon fossa which resulted from periosteal stripping at the time of injury. This tissue must be removed completely. Excision of scar tissue must be adequate and must be carried out until both trochlea and capitellum are cleared, whereupon reduction can be effected. The capsule is sutured. Then the joint is carried through the full range of motion. If flexion is limited, this may be due to an obstacle anteriorly (bony prominence of the coronoid, ossification in the brachialis, scar tissue in coronoid fossa) or insufficient freeing of capsule posteriorly. When flexion is deemed to be adequate, the triceps aponeurosis may be found to have displaced distally in relation to its muscle fibers. Therefore, after approxi-

[7] King, T: Recurrent dislocation of the elbow, J. Bone & Joint Surg. 35B:50, 1953

mating the muscle, the aponeurosis is sutured in place while the elbow is held flexed. Otherwise shortening of the triceps mechanism will shorten the arc of flexion.

RUPTURE OF THE BICEPS INSERTION AT THE ELBOW

The biceps tendon may be ruptured or avulsed at or near its insertion to the bicipital tuberosity of the radius. The usual mechanism of injury is that of forcible flexion of the elbow against strong resistance. An example is that of a worker assisting in lifting a heavy object and suddenly bearing the full weight of that object when the fellow worker lets it slip from his grasp. A painful snap or tearing sensation is experienced at the elbow, followed immediately by weakness of active flexion of the elbow and supination of the forearm. The antecubital space is tender, but swelling is minimal because of the tense overlying fascia. On attempting active flexion, a bulbous swelling forms in the upper arm, the retracted belly of the biceps.

Pathologically, rupture is preceded by degenerative changes in the tendon, often associated with advanced age and arteriosclerosis. The bicipital tuberosity often presents an irregular jagged ridge which conceivably erodes the tendon and causes it to fray and rupture.[8] If the tendon is partially torn but not as yet completely avulsed, it may produce symptoms of localized pain on attempting motions requiring lifting and supination. Tenderness and a sensation of fine crepitus may be palpable over the tuberosity.

Treatment

The tendon is recovered and sutured to the brachialis at its insertion This results in restoration of strong flexion, but supination remains weak. When the occupation of the patient demands good rotatory power of the forearm, an attempt must be made to resuture the tendon to the bicipital tuberosity.

Technic. A longitudinal incision is made anteromedially over the lower third of the arm, then carried transversely across the front of the elbow and then a little distally along the lateral aspect of the forearm. The tendon is secured in the upper arm and brought down. The radial recurrent vessels are ligated and cut, and the main vessels and the radial and the ulnar nerves are retracted medially. The forearm is strongly supinated, and the tuberosity of the radius is palpated and exposed by subperiosteal dissection. A drill hole is made anteroposteriorly through the tuberosity, and the tendon is drawn through and sutured to itself. Postoperatively, the elbow is immobilized in flexion for approximately 6 weeks before exercises are instituted.

TUBERCULOSIS OF THE ELBOW[9]

Osseous tuberculosis of the elbow like tuberculosis of other bony structures may be mild or severe, depending upon the intensity of the allergic exudative response. This acute early exudative inflammation produces the clinical picture of pain, muscle spasm, generalized joint tenderness, swelling which consists of boggy edema of the periarticular tissues and increased synovial effusion; also, marked limitation of motion. The acute stage then resolves either completely or incompletely with the formation of caseous destructive lesions which slowly enter the joint, destroy the intra-articular tissues and may heal at this point by a fibrous ankylosis, rarely by bony ankylosis. Or the caseous destructive arthritis may continue indefinitely with formation of persistent draining sinuses and loss of all joint motion. (See the discussion on etiology and pathology under Tuberculosis in section on Infections.)

Types of Lesions

The *acute exudative lesion* is an acute inflammation of bone (or synovium) which causes an extreme degree of diffuse osteoporosis of the involved bone. This condition may subside with complete restoration of joint motion. Or this inflammation may subside to a low grade but predominantly *destructive caseous lesion* which is seen as an irregular cavitation in the bone. When the destructive lesion is located in the coronoid process of the ulna, the joint eventually and invariably becomes infected. When destruction is at a dis-

[8] Davis, W M., and Yassine, Z · An etiological factor in tear of the distal tendon of the biceps brachii, J. Bone & Joint Surg 38A:1365, 1956.

[9] Wilson, J. N.: Tuberculosis of the elbow, J Bone & Joint Surg. 35B.551, 1953.

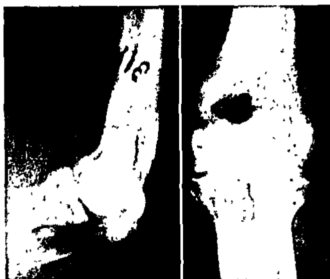


FIG 321. Tuberculosis of the elbow. (N U. Case No 284)

tance from the joint, e.g , in the medial or the lateral epicondyle, the outlook is more favorable The destruction may become massive by spread from the joint to all contiguous structures, all semblance of a joint is lost, and the head of the radius subluxates or dislocates posteriorly.

COMPLICATIONS

Sinuses occur only when actual bony destruction is present The ulnar nerve may be compressed or engulfed in cicatrix.

TREATMENT

When roentgenograms reveal the diffuse osteoporosis of the exudative reaction, conservative treatment by prolonged immobilization plus antibiotics may effect a cure. A localized destructive lesion, particularly of the coronoid, demands urgent surgical eradication. It is wise to excise the focus to eradicate a source of future exacerbations with extension to the joint. When the lesion is in the coronoid or destruction is extensive, arthrodesis is necessary. The position of immobilization depends upon the final desired function. Ankylosis in a position of 120° (30° below the right angle) presents the best cosmetic result. One can type, reach into the hind pocket, and the arm hangs naturally at the side. If the function of feeding is necessary, the right angle position is best

Posterior dislocation of the radius is treated by excision of the radial head.

Arthrodesis of the Elbow. A posterior midline incision is made over the lower end of the humerus and extending over the olecranon. The ulnar nerve is isolated from its groove between the olecranon and the medial epicondyle and is retracted. The flat triceps tendon is severed at the musculotendinous junction, and the flap remaining attached to the olecranon is retracted downward, exposing the posterior joint capsule. The capsule is cut longitudinally, and the soft tissues are elevated from the medial and the lateral aspects of the joint and retracted forward, giving access to the anterior regions. The elbow is strongly flexed, thereby exposing the articular cartilage of radius, ulna and humerus. All cartilage, granulation tissue, abnormal appearing bone and thickened capsule are removed until normal cancellous bony structures are approximated. If the radial head is subluxated, it is removed. A hole is drilled into the coronoid fossa to receive a tibial cortical graft, which is laid into a gutter cut in the posterior surface of the humerus. Catgut sutures aid in

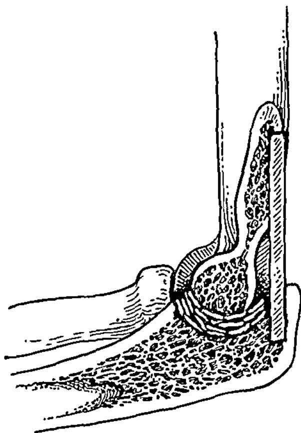


FIG. 322. Arthrodesis of the elbow. Bone chips fill the humero-ulnar interval. A strong cortical graft provides fixation.

holding the graft in place. The joint space is packed with cancellous bone chips. The tendinous flap is sutured to the muscle, and the ulnar nerve is restored to its bed. A cast is applied either at or below a right-angle position.

OSTEOCHONDRITIS DISSECANS OF THE ELBOW[10]

This condition is analogous to that occurring more frequently in the knee joint. A small segment of subchondral bone slowly separates from its bed. The overlying articular cartilage, at first intact, also slowly separates, and an osseocartilaginous body is extruded into the joint cavity. The incidence in the elbow is second only to that of the knee, which is affected in 85 per cent of cases.

Etiology

The condition becomes apparent in adolescence, and males are preponderantly affected. The right elbow is involved most often; occasionally, the condition is bilateral.

Repeated minor injuries have been blamed, but this is questionable in view of the fact that pneumatic drill workers display no greater than the usual incidence of osteochondritis dissecans.[11]

Pathology

The features are similar to osteochondritis dissecans occurring elsewhere. Early, the overlying intact articular cartilage appears normal, or slight discoloration and fibrillations indicate the site of underlying pathology. Later, the surface of the capitellum is ragged, and a flap of cartilage may overhang a deep crater. One or several loose bodies may be found within the joint cavity, especially anteriorly or posteriorly in the supratrochlear fossa. The largest loose body consists of a center of bone and a covering of hyaline cartilage. The smaller ones consist entirely of cartilage and therefore are not visible in roentgenograms. (For microscopic appearance, see Osteochondritis Dissecans in section on the Knee.)

The following features are peculiar to the lesion of the elbow:

1. *Enlargement of the head of the radius* characteristically in a posterolateral direction. The metaphysis is often funnel-shaped.

2. *Epiphysial prematurity* or premature fusion of the epiphyseal lines.[12] Fusion always begins on the lateral aspect and proceeds medially.

Clinical Picture

The onset may be insidious, and the first indication of trouble is a diffuse, dull ache accompanied by a little stiffness. A history of trauma is often obtained, but its causative relationship is questionable.

Often the elbow is asymptomatic until a loose body produces a dramatically sudden onset of spontaneous locking and pain followed by effusion.

The condition may be entirely silent and discovered accidentally.

The most constant signs are limitation of extension of the elbow and palpable enlargement of the head of the radius.

Roentgenologic Findings

The lesion is most frequently found in the capitellum, infrequently in the head of the radius, rarely in both. The typical appearance of the lesion observed in the knee, the hip and the ankle, i e., an island of subchondral bone surrounded by a zone of rarefaction, is seldom seen in the elbow.

1. **Early Changes.** A patchy rarefaction with ill-defined limits largely affecting the convexity of the capitellum, usually best seen in A-P views. The rarefaction becomes pronounced, multiple small fragments separate. and a ragged appearance is produced. Eventually, a single large punched-out cavity may result.

The head of the radius, when the major change is in the capitellum, becomes irregular. When the lesion is confined to the radial head, the medial portion of the convex rim displays the typical changes ordinarily found in the knee, i.e., an island of subchondral bone surrounded by a zone of rarefaction.

2. **Late Changes.** Loose bodies of bony density may be apparent, but, when not evi-

[10] Roberts, R., and Hughes, R: Osteochondritis dissecans of the elbow joint, J. Bone & Joint Surg 32B 348, 1950

[11] Rostock, P: Arch orthop u. Unfall-Chir 33: 449, 1933

[12] Löhr, W: Arch. klin Chir. 157:752, 1929; 162:489, 1930.

dent, they are composed solely of hyaline cartilage. *Flattening of the capitellum* is a constant feature. The punched-out defect or irregular eburnated margins indicate the site of the original lesion and the source of the loose body. A large loose body is seen not infrequently in the supratrochlear fossa and appears to originate from the supratrochlear septum. However, no cartilage exists at this site, and careful examination invariably reveals the source as elsewhere.

The late sequel is observed in the form of degenerative arthritic changes appearing first at the radiocapitellar articulation and spreading to the remainder of the joint. The radial head is practically always enlarged.

Differential Diagnosis

The condition may be confused with osteochondritis juvenilis of the capitellum. The latter is characterized by bilateral, irregular ossification that simulates irregularity of the capitellum. However, the capitellum is composed mainly of cartilage and is normal in external appearance Eventually, one observes in roentgenograms complete reossification and restitution of the bony process. The condition usually occurs in childhood, causes no symptoms, and results in little or no disability. Loose bodies never form.

Treatment

Operation is not advisable in the early case. Rest of the elbow in a sling over a prolonged period of time will often effect a cure as indicated in roentgenograms by restoration of bony architecture.

Surgery is indicated for late cases with incidents of locking and beginning degenerative arthritic changes. Loose bodies should be removed. The irregular articulation betwee radial head and capitellum by erosion c tinues to shear off fragments of cartil which, lying free within the joint, continue grow and enlarge. Therefore, removal of the radial head is necessary to prevent recurrence of loose bodies and to halt spread of degenerative changes. It is also indicated to improve motion.

MYOSITIS OSSIFICANS IN THE ELBOW

Myositis ossificans is prone to develop about the elbow following injuries, particularly posterior dislocation and supracondylar fracture of the humerus. The abnormal heterotopic ossification develops within the lower end of the brachialis anticus, suggesting that the periosteum has been stripped up at the insertion of the muscle. The process is more likely to follow if the elbow is manipulated forcefully. In the early stages of development, all forms of excessive movement and manipulation should be avoided. No attempt should be made to operate upon the joint, and the mass must not be excised, because the trauma of operation will greatly aggravate the process, causing it to extend and rendering the joint hopelessly beyond repair. The ossification, if left alone, will increase for an indefinite period of time, then becomes stationary, and finally regresses, becomes solid and well-defined and at times may disappear completely. If the final bony mass does not alter its appearance over many months, and if it interferes with joint function, it may be removed. (See Myositis Ossificans in section on Diseases of Muscle.)

23

The Hand

SURGICAL ANATOMY*

Skin and Subcutaneous Fascia

The *palmar aponeurosis* is firmly fixed to the overlying skin by fibrous bands which extend into the subcutaneous fat. This fibrous tissue hypertrophies in Dupuytren's contracture. The dorsal skin is lax and lies on loose areolar tissue which forms the *dorsal subcutaneous space* through which pass veins, lymphatics and sensory nerves. This space receives most of the lymph flow from the palmar aspect of the hand and explains the marked edema of the dorsum from infection of the palm.

The *dorsal deep fascia* fuses with the extensor tendons, forming the roof of the *dorsal subaponeurotic space.* Infection here is rare except by penetration or extension from osteomyelitis of a metacarpal.

The Palmar Deep Fascia

That portion which covers the thenar and the hypothenar muscles is intimately attached to these muscles so that no space exists. The central triangular portion, the *palmar aponeurosis,* is continuous proximally with the palmaris longus tendon, which assists in flexion of the hand. The majority of hand infections are beneath this. The aponeurosis sends extensions distally to fuse with the fibrous tendon sheaths over the proximal phalanges. These aponeurotic prolongations in the distal part of the palm are connected with each other by transversely disposed fasciculi which cover the digital vessels and nerves and form the *superficial transverse metacarpal ligament.*

In the middle of the palm, a septum extends deeply from the aponeurosis to the third metacarpal, separating the thenar space from the midpalmar space. Distally, septa extend from the deep aspect of the palmar aponeurosis to form annular fibrous canals for the passage of flexor tendons, lumbrical muscles, digital vessels and digital nerves. Each pair of septa passing deeply to attach to a metacarpal form a canal for passage of the corresponding tendons to that finger. Between these tendinous passages run the lumbrical muscles, the digital vessels and the digital nerves. When carrying out a dissection for removal of the aponeurosis in Dupuytren's contracture, it is essential first to isolate the small vital structures in the intertendinous compartments before resecting the fascia.

Blood and Lymph Vessels

The *radial* and the *ulnar* arteries each terminate in a superficial branch and a deep branch. The superficial branches form the *superficial palmar arch,* and the deep branches the *deep palmar arch.* A line drawn across the palm at the level of the distal border of the fully abducted thumb marks the location of the superficial palmar arch. A finger's breadth proximal to this lies the deep arch. The pulsations of the ulnar artery are felt just radial to the pisiform bone. At the level of the wrist, the ulnar artery gives off volar and dorsal carpal branches to form with similar branches from the radial artery an arterial wristlet about the carpal bones.

The deep palmar arch sends perforating branches between the proximal ends of the metacarpals to connect with the dorsal carpal arch. The latter sends small branches distally to the phalanges. Metacarpal branches from the deep arch pass distally to empty into digital branches of the superficial arch proximal to their bifurcation.

Pulsations of the radial artery are palpated near the proximal volar carpal skin crease. Its superficial branch contributes to the super-

* Lampe, E. W.: Surgical Anatomy, Ciba Clinical Symposia 9 3-45 (No. 1), 1957.

ficial arch. The larger deeper branch passes under the "snuffbox" tendons and plunges between the two heads of the first dorsal interosseous muscle to reach the palm where it forms the greater part of the deep palmar arch. The superficial arch gives off digital branches which bifurcate into phalangeal branches immediately deep to the palmar aponeurosis. The arch and the digital branches lie superficial to branches of the median and the ulnar nerves, but the relationship is reversed in the fingers.

Importance of the Osseous Blood Supply. The lunate bone receives branches from the volar and the dorsal carpal arteries via the volar and the dorsal ligaments which attach to the radius. When the lunate is dislocated and one ligament is torn, the bone survives. If both ligaments are torn, the blood supply is interrupted, and aseptic necrosis is the result.

In the navicular bone, multiple nutrient vessels provide adequate nutrition to the entire bone. However, in one third of cases, the greatest concentration of blood vessels is disposed toward the distal portion of the bone; consequently, a fracture will interrupt the blood supply to the proximal pole.

When infection occurs at the distal end of the finger, the exudate is enclosed by rigid fibrous septa. Accumulation of exudate within unyielding compartments compresses nutrient vessels and causes necrosis of the bony tuft.

Veins. The *dorsal venous arch* on the dorsum of the hand receives digital veins and drains into the *cephalic and basilic veins* on the radial and ulnar borders of the wrist. The small superficial palmar veins empty into the median antebrachial vein. The *deep system* is associated with arteries as venae comites. These unite at the elbow to form the *brachial vein*, which unites with the basilic vein at the pectoral fold to form the axillary vein. The superficial and the deep veins anastomose throughout the extremity by communicating veins.

Lymph Vessels. These follow the veins. Most of the lymph from the palm and the fingers flows into the loose areolar area of the dorsal subcutaneous space and follows the vessels accompanying the cephalic and the basilic veins. A lymph gland in the deltopectoral interval is the first sizable gland encountered in the pathway along the cephalic vein, and this is the most direct route to the thoracic duct. In contrast, the route along the basilic and the axillary veins is gland-studded. The deep palmar lymph vessels follow along the superficial and the deep palmar arches, then continue along the venae comites of the radial and the ulnar arteries.

Nerves (Plate 46)

Motor. The *ulnar nerve* supplies the flexor carpi ulnaris and the ulnar half of the flexor digitorum profundus. The remainder of the flexor-pronator group (flexor carpi radialis, flexor digitorum sublimis, radial half of the flexor profundus, pronator teres, pronator quadratus) is controlled by the *median nerve.* The *radial nerve* supplies the extensor-supinator group of muscles (extensor carpi ulnaris, extensor carpi radialis longus and brevis, extensor digitorum communis, extensor indicis proprius, extensor digiti quinti proprius, extensors pollicis longus and brevis, abductor pollicis longus, supinator longus). The ulnar nerve supplies all the intrinsic muscles except those of the thenar eminence and the two adjacent lumbrical muscles which are controlled by the median.

To test for the ulnar nerve, spreading and approximating the fingers will indicate normal function of the interossei. The median nerve is tested by performing the motion of the opponens muscle, i.e., apposing the tip of the thumb to the tip of each finger.

Sensory. The *median nerve* supplies the palmar aspect of the thumb, the index, the middle and the radial half of the ring fingers; the corresponding portion of the palm; and the dorsum of the distal 2 phalanges of the lateral 3½ fingers.

The *ulnar nerve* supplies the volar and the dorsal aspects of the little finger and the ulnar half of the ring finger and the corresponding portion of the palm and the dorsum of the hand.

The *radial nerve* supplies the lateral two thirds of the dorsum of the hand, a portion of the thenar eminence, and the dorsum of the proximal phalanges of the lateral 3½ fingers.

The *median nerve* passes under the rigid transverse carpal ligament (Plates 46 and 47) and divides into (A) *3 lateral branches,* 2 supplying either side of the thumb and the

other the lateral side of the index finger, and (B) *2 medial branches,* one dividing to innervate adjacent surfaces of the index and the middle fingers and the other to adjacent aspects of the 3rd and the 4th fingers.

It is important to remember that *the main muscular branch of the median nerve in the hand arises from the lateral cutaneous branch to the thumb just distal to the transverse carpal ligament and passes over the flexor pollicis longus tendon and its sheath. Therefore, any incision skirting the thenar eminence or incising the tendon sheath of the flexor pollicis longus should not extend further proximally than the level of the midpoint of the first metacarpal bone.*

From nerves supplying the lateral aspect of the index finger and adjacent sides of the 2nd and the 3rd fingers arise small branches for the second and the third lumbrical muscles, respectively.

The *ulnar nerve* passes lateral to the pisiform bone and bifurcates into (A) a *superficial ramus* which supplies filaments to the palmaris brevis, a branch to the medial aspect of the little finger, a branch to apposing aspects of the little and the ring fingers, and a twig connecting with the medial branch of the median; and a (B) *deep ramus* which supplies muscular branches to the hypothenar eminence (abductor, flexor and opponens digiti quinti), the volar and the dorsal interossei, the adductor pollicis, the lumbrical muscles 3 and 4, and the deep head of the flexor pollicis brevis.

Muscles. Muscles of the hand can be grouped as *extrinsic* and *intrinsic.* The muscle bellies of the former are located in the forearm, but their tendons play an important part in movements of the hand.

SUPINATION is controlled by the *biceps,* which inserts into the tuberosity of the radius, and the *supinator* muscle which extends from the dorsolateral aspect of the ulna and wraps itself laterally around the proximal fourth of the radius. The musculocutaneous nerve innervates the biceps, while the radial nerve supplies the supinator.

PRONATION is controlled by the *pronator teres* and the *pronator quadratus,* both supplied by the median nerve.

ADDUCTION of the hand at the wrist is produced by the combined action of the *flexor carpi ulnaris* and the *extensor carpi ulnaris* muscles, the former innervated by the ulnar nerve and the latter by the radial. The ulnar flexor inserts into the pisiform, the hamate and the base of the 5th metacarpal. The ulnar extensor inserts into the base of the 5th metacarpal bone.

ABDUCTION is performed by synergistic action of the *flexor carpi radialis* and the *extensors carpi radialis longus and brevis.* All these muscles insert upon the base of the 2nd and the 3rd metacarpal bones.

The carpal flexors and extensors, in addition to their individual function, work together to stabilize the hand upon the forearm. Particularly important is their action of holding the hand in dorsiflexion, relaxing the finger extensors and permitting maximum efficiency for finger flexors.

FLEXION OF THE FINGERS is produced mainly by the *flexors digitorum sublimis and profundus.* The assistant flexor role of the lumbricals and interossei will be discussed under the description of intrinsic muscles.

The *flexor digitorum sublimis* originates from the medial humeral epicondyle, the coronoid process of the ulna, and the proximal two thirds of the volar margin of the radius. Proximal to the volar carpal ligament, the muscle gives rise to 4 tendons inserting into the proximal thirds of the middle phalanges of the medial 4 digits. The vincula longa and breva give additional insertion into the proximal end of the middle phalanges.

The following surgical points must be remembered:

1. Beneath the volar carpal ligament, *the 3rd and the 4th sublimis tendons lie superficial* to the 2nd and the 5th tendons. Occasionally, the 3rd, the 4th and the 5th tendons will lie superficially in the same plane while the tendon of the index finger lies behind the middle finger tendon. Deeply, the profundus tendons all lie in one plane (Plate 47).

2. The *insertion of the sublimis* tendon is peculiar. The tendon first splits at the proximal end of the first phalanx to permit passage of the profundus tendon and then reunites, only to split a second time to gain insertion on each side of the proximal third of the middle phalanx (Plates 48 and 49).

The *flexor digitorum profundus* originates from the proximal two thirds of the ulna and the adjacent interosseous membrane. It inserts into the base of the distal phalanx (Plate 49) of the medial 4 digits. The vincula offer additional insertion.

Beneath the transverse carpal ligament, it is not uncommon to see only one profundus tendon to the index finger while the remainder of the profundus is fibromuscular. The latter portion, about 2 cm. distal to the transverse carpal ligament, divides into tendons to the 3rd, the 4th and the 5th fingers.

EXTENSION OF THE FINGERS AND THE THUMB is produced by the combined action of extrinsic and intrinsic muscles.

The *extensor digitorum communis* originates from the lateral humeral epicondyle, the intermuscular septum and the antebrachial fascia. It divides into 4 tendons which pass through the fourth compartment of the dorsal carpal ligament at the wrist. It is not uncommon to observe only 3 tendons passing through the 4th dorsal compartment. In this instance, the 4th tendon arises on the dorsum of the hand from the 4th finger extensor and goes from there to the 5th finger. The *extensor indices proprius* also passes through this compartment.

Each tendon of the extensor digitorum communis (Plate 49) inserts into the dorsum of the base of a proximal phalanx where (1) it extends the proximal phalanx, and (2) stabilizes the proximal phalanx in extension so that the intrinsic muscles can extend the middle and the distal phalanges and impart lateral motion to the fingers.

The extensor communis tendon then continues distally to contribute to the formation of the dorsal aponeurosis. It divides into 3 parts. The central slip inserts into the dorsum of the base of the middle phalanx. Two lateral tendinous slips unite with tendons of the lumbrical and the interosseous muscles to form a lateral band on each side which extends forward to insert on the dorsum of the base of the distal phalanx.

The action of the extensor digitorum communis is mainly to extend the proximal phalanx. With the finger in full flexion, it also acts to extend the distal 2 phalanges However, as the finger slowly extends, the force of extension becomes progressively less, and the extensor movement is gradually taken over by the intrinsics via the lateral bands. During the last 45° of extension, extension of the distal 2 phalanges by the extensor communis is negligible and is accomplished almost exclusively by the intrinsics. Because the extensor communis tendon is inserted into the proximal phalanx, when the muscle contracts, practically all of the tension is taken up in extending the proximal phalanx. It is while the proximal phalanx is extended that the intrinsics exert their maximum effort in extending the middle and the distal phalanges.

However, the situation changes when the extensor digitorum communis relaxes sufficiently to permit the flexors digitorum sublimis and profundus to begin flexing the middle and the distal phalanges. The "hood" which overlies the metacarpophalangeal joint and into which the intrinsics attach shifts forward and comes to rest over the proximal phalanx. In this position, contraction of the lumbricals and the interossei effect tension upon the "hood" of the extensor apparatus and then flex the proximal phalanges. Thus the important flexors of the proximal phalanges are the intrinsics.

The *extensor digiti quinti* passes through the 5th compartment under the dorsal carpal ligament. It arises by a thin tendinous slip from the common extensor in the forearm and it inserts into the dorsal aponeurotic hood of the 5th finger. It aids in extending the proximal phalanx.

The tendon of the *extensor indicis proprius* passes through the 4th dorsal compartment with the extensor communis tendons It arises from the ulna at the junction of the proximal three fourths with the distal fourth of the bone, and its function is to assist extension of the proximal phalanx of the index finger.

The *abductor pollicis longus* arises by 3 heads of origin from the proximal part of the ulna, the interosseous membrane, and the middle third of the radius Its tendon passes through the first dorsal compartment over the lateral aspect of the radius (Plates 50 and 51). A stenosing tenosynovitis at this point (de Quervain's disease) causes great disability of the thumb. The tendon inserts, often by as many as 3 slips, to the dorsum of the base of

the very mobile first metacarpal. This muscle abducts the first metacarpal and acts to stabilize the first carpometacarpal joint in the act of thumb pinch. Loss of this muscle causes severe disability of the thumb.

The *extensor pollicis brevis* arises from the interosseous membrane and the dorsum of the radius just distal to the abductor pollicis longus, and its tendon passes through the first dorsal compartment in company with the long abductor; it, too, may be involved by a stenosing tenosynovitis (Plates 50 and 51). It inserts into the dorsum of the base of the proximal phalanx of the thumb, extending this phalanx and aiding in extending and abducting the hand at the wrist.

The *extensor pollicis longus* is larger than the brevis. It arises from the middle third of the ulna and the interosseous membrane, and its tendon passes through the third dorsal compartment (Plates 50 and 51). In this latter situation, it may be severed by a fragment of a Colles' fracture, or by friction may rupture 5 or 6 weeks after a fracture. The tendon inserts into the dorsum of the base of the distal phalanx of the thumb, extending this phalanx and assisting in extending the hand at the wrist.

Arrangement of Tendons, Vessels and Nerves at the Wrist (Plates 47 and 51)

By clenching the fist tightly and flexing the wrist against resistance, the tendon of the palmaris longus is rendered prominent and may be utilized as an orientation point. However, it must be remembered that the palmaris longus is absent in 10 per cent of cases. The tendon runs superficial to the transverse carpal ligament. Behind the tendon lies the median nerve beneath the ligament. Radialward from the palmaris longus may be felt the stout flexor carpi radialis. The latter lies between the radial artery laterally and the flexor pollicis longus medially and lying deeply.

Over the ulnar aspect, the flexor carpi ulnaris is prominent. Immediately radial to this tendon lie the ulnar nerve and the ulnar artery. The 2 flexor tendon quartets lie just medial to the centrally placed palmaris longus tendon. Those acting on the 3rd and the 4th fingers lie superficial to those of the 2nd and the 5th fingers. This can be remembered by placing one's 2nd and 5th fingers behind the 3rd and the 4th fingers. Not uncommonly, the sublimis tendons to the 3rd, the 4th and the 5th fingers lie in the superficial plane, and the index finger sublimis tendon lies behind that of the middle finger.

The 4 flexor digitorum profundus tendons lie in one plane behind the sublimis tendons. It is not unusual at this point to observe only the profundus tendon to the index finger while the remainder of the structure is fibromuscular—the other 3 tendons arising 1 or 2 cm. more distally. Beneath the profundus tendons lies the pronator quadratus muscle. A *subtendinous space* (Parona) exists between the profundus tendons and the pronator quadratus. This space is important as a possible site of infection.

By insular attachments on the dorsal and the lateral aspects of the radius and the ulna, the dorsal carpal ligament creates 6 compartments for the passage of extensor and abductor tendons.

Over the radial and dorsal aspect of the wrist, the *anatomic snuffbox* is formed by the 3 thumb tendons (Plate 50). The volar boundary of the snuffbox is formed by the abductor pollicis longus and the extensor pollicis brevis tendons which pass through the 1st compartment. The dorsal boundary is formed by the tendon of the extensor pollicis longus passing through the 3rd compartment. Between the thumb tendons and lying deeply are the 2 radial extensor tendons in the 2nd compartment. The radial artery runs through the snuffbox on its way to the first interosseous space. Deep to the radial artery is the capsule of the wrist joint and the navicular (scaphoid) bone The sensory branches of the radial nerve are also found in the snuffbox.

The 4 communis tendons can easily be associated with the 4th dorsal compartment, the extensor indicis proprius tendon accompanying these tendons. When, as is often the case, the 4th communis tendon arises distally in the dorsum of the hand, the 4th dorsal compartment will be occupied by only 3 extensor communis tendons and the extensor indicis proprius tendon.

Intrinsic Muscles of the Hand (Plates 49 and 52)

These may be grouped into those forming the hypothenar eminence, the thenar eminence, and a third group composed of muscles situated between these two eminences.

Muscles of the Hypothenar Eminence (Plate 52). The abductor digiti quinti, the flexor digiti quinti brevis and the opponens digiti quinti comprise this group. The *abductor digiti quinti* arises from the pisiform and the pisohamate ligament and inserts into the capsule of the 5th metacarpophalangeal joint, the ulnar aspect of the base of the proximal phalanx, and the ulnar border of the extensor aponeurosis of the 5th finger. It abducts the 5th finger when the proximal phalanx is extended and flexes the proximal phalanx when the long extensor is relaxed.

The *flexor digiti quinti brevis* arises from the hamate bone and the transverse carpal ligament and is inserted into the ulnar side of the base of the 1st phalanx. It assists in abducting the 5th finger and in flexing the proximal phalanx.

The *opponens digiti quinti* originates from the hamate bone and the transverse carpal ligament and is inserted into the ulnar border of the 5th metacarpal bone. Because the 5th is the second most mobile of the metacarpals, the opponens is able to draw the 5th metacarpal forward, thereby helping to deepen the hollow of the palm. The unimportant palmaris brevis merely corrugates the skin on the ulnar side of the palm.

All muscles of the hypothenar eminence are innervated by the ulnar nerve.

Muscles of the Thenar Eminence (Plate 52). This group consists of the abductor pollicis brevis, the flexor pollicis brevis and the opponens pollicis muscles, all supplied by the median nerve.

The *abductor pollicis brevis* arises from the transverse carpal ligament and the navicular and the greater multangular bones. It inserts into the radial side of the proximal phalanx of the thumb. It aids in abducting the thumb away from the palm.

The *opponens pollicis* originates from the transverse carpal ligament and the greater multangular bone. It inserts into the whole radial side of the first metacarpal bone. Its function is to rotate the thumb and bring it forward into the palm so as to approximate the tip of the palm to the tips of the fingers. The *thumb-finger approximator test* is used to test for the integrity of the median nerve.

The *flexor pollicis brevis* consists of 2 parts: (1) a *superficial portion*, innervated by the median nerve, originating from the transverse carpal ligament and the greater multangular bone and inserting into the radial side of the proximal phalanx; and (2) a *deep portion*, innervated by the ulnar nerve, arising from the ulnar aspect of the 1st metacarpal and inserting into the ulnar aspect of the base of the proximal phalanx.

Interosseous Muscles. There are 7 interossei: 4 in the dorsal group and 3 in the volar group.

The *4 dorsal interossei* (Plate 52) are bipennate with their muscular heads of origin from adjacent sides of the metacarpal bones. One head of each muscle inserts into the tubercle on the lateral aspect of the base of the proximal phalanx, or into the capsule of the M-P joint, or into both tubercle and capsule. The other head inserts into the lateral band of the dorsal aponeurosis through which it acts to extend the distal 2 phalanges. Often this latter tendon sends a slip to the capsule of the M-P joint. When the proximal phalanx is fully extended, contraction of the muscle belly inserting upon the tubercle will cause lateral motion of the finger; contraction of the other muscle belly aids in extending the middle and the distal phalanges. When the extensor digitorum communis relaxes, the aponeurotic hood over the M-P joint, to which the interosseous sends fibers, shifts forward over the proximal phalanx, and the interosseous then flexes the proximal phalanx.

The 2nd and the 3rd dorsal interosseous muscles insert into the tubercles on the radial and the ulnar sides of the proximal phalanx of the middle finger, respectively, and into the lateral bands of the dorsal expansion hood This enables the muscles to stabilize the middle finger while the index, the ring and the little fingers can be abducted away from the middle finger. This is accomplished because the first dorsal interosseous inserts into the tubercle on the radial side, and the 4th dorsal interosseous and the abductor digiti quinti

insert into the tubercle on the ulnar side of their respective proximal phalanges.

The *volar interossei* (Plate 52) are 3 in number. The first arises from the ulnar side of the volar aspect of the 2nd metacarpal bone and is inserted into the same side of the proximal end of the 1st phalanx and the lateral band of the dorsal aponeurosis. The 2nd and the 3rd arise from the radial side of the 4th and the 5th metacarpal bones, respectively, and are inserted into the same side of the proximal phalanx of the 4th and the 5th fingers and the lateral bands. Contraction of these muscles approximates the 2nd, the 4th and the 5th fingers toward the middle finger. In addition, by virtue of their insertion into the lateral bands and the hood over the proximal phalanx, they assist other intrinsic muscles in flexing the proximal phalanx and in extending the middle and the distal phalanges.

The *adductor pollicis muscle* functionally is associated with the volar interossei and similarly is innervated by the ulnar nerve. The *oblique head* of the adductor pollicis arises from the capitate bone and the bases of the 2nd and the 3rd metacarpals. The *transverse head* arises from the distal two thirds of the volar aspect of the 3rd metacarpal bone. Most of the fibers of the oblique head and all of the transverse head are inserted into the ulnar side of the base of the proximal phalanx of the thumb. Not uncommonly, a sesamoid bone is found in this tendon. Some of the fibers from the oblique head join the flexor pollicis brevis to insert into the radial side of the thumb's first phalanx. This tendon also contains a sesamoid bone. The tendon of the flexor pollicis longus lies between these two points of insertion.

As the name implies, the function of the adductor pollicis muscle is adduction of the thumb toward the third metacarpal. This contrasts with the action of the opponens muscle, which is controlled by the median nerve. The latter swings the thumb in an arching manner toward the tips of the fingers, whereas the former slides or scrapes the thumb across the palm.

An important function of the adductor pollicis is to aid the flexor pollicis brevis in stabilizing the metacarpophalangeal joint in flexion when performing a pinching action between the thumb and the index finger. Loss of these muscles in ulnar nerve paralysis will cause the metacarpophalangeal joint to fall into hyperextension as pinch is weakly attempted.

Lumbrical Muscles (Plates 48 and 49). On the radial side of the palmar portion of each flexor digitorum profundus tendon is a lumbrical (wormlike) muscle. The 1st and the 2nd lumbricals originate respectively from the radial side of the 1st and the 2nd flexor profundus tendons; the 3rd lumbrical originates from the adjacent sides of the 2nd and the 3rd profundus tendons; and the 4th from the adjacent sides of the 3rd and the 4th profundus tendons. Each muscle sends a tendon along the radial side of the M-P joint. The tendon sends one slip into the dorsal hood which shifts over the M-P joint and the proximal phalanx; and another slip fuses with a contribution from the interosseous muscles to form a lateral band. When the extensor digitorum communis contracts to extend the proximal phalanx, the hood is drawn proximally, and the force of contracting intrinsics is expended along the lateral band to extend the distal 2 phalanges. When the extensor digitorum communis relaxes, the dorsal hood shifts forward over the proximal phalanx, and the force of contracting intrinsics is expended through the hood upon the proximal phalanx, and flexion of the latter takes place.

The first 2, or lateral, lumbricals are innervated by the median nerve; the 3rd and the 4th, or medial, lumbricals are innervated by the ulnar nerve. This corresponds to a similar innervation of the profundus muscle.

Ligaments of the Hand

A thick but loose articular capsule holds together the saddle-shaped joint between the 1st metacarpal and the greater multangular bone. The configuration of the facets and the loose capsule permits a wide range of movement at this joint.

The bases of the 2nd, the 3rd, the 4th and the 5th metacarpals are held together by dorsal, volar and interosseous ligaments.

The volar surfaces of the 2nd to the 5th metacarpal heads are connected by a tough fibrous band, the *deep transverse metacarpal ligament* (Plate 52). This ligament and the

ligaments about the base of the metacarpals give the hand stability. They permit the 5th metacarpal a 30° range of movement; the 4th about 15°. They permit practically no mobility to the 2nd and the 3rd metacarpal bones, thereby making this the most stable part of the hand. The deep transverse metacarpal ligament helps to preserve the metacarpal arch, and its rupture weakens the hand to a marked degree. The lumbrical tendons course over its palmar aspect while the interosseous tendons run over its dorsum.

The accessory volar and 2 collateral ligaments strengthen the metacarpophalangeal joint. The collateral ligaments are particularly important. When the M-P joint is fully extended, the collateral ligaments are redundant and permit mediolateral movement of the finger. When the joint is flexed, the ligaments must encircle the wider volar aspect of the metatarsal head and, therefore, are tightened up. When the collateral ligaments are allowed to shorten, e.g., by prolonged immobilization of the joint in extension, flexion will be prevented because of inability of the ligaments to ride over the condyles.

Tendon and Muscle Sheaths of the Hand (Plates 47, 48, 51)

The tendon sheaths of the 2nd, the 3rd and the 4th fingers extend from the terminal phalanges to the distal end of the palm approximately to a line drawn across the palm from the medial end of the distal palmar crease to the lateral end of the proximal crease. The proximal ends of these sheaths overlie the distal ends of the thenar and the midpalmar spaces or bursae, thereby explaining spread of infection from sheaths of these fingers to these spaces. Rarely, any one of these sheaths may extend proximally as far as the wrist.

Kanavel described 4 cardinal points for diagnosing the presence of pus in flexor tendon sheaths:

1. *Finger held in flexion* to relieve tension.
2. *Finger is uniformly swollen.*
3. *Intense pain on passive extension of finger.*
4. *Tenderness along course of tendon sheath.*

The flexor sheath of the thumb usually extends from the terminal phalanx to a point 2 or 3 cm. proximal to the proximal volar crease of the wrist. The proximal half is somewhat redundant and is commonly referred to as the *radial bursa.* Occasionally, the radial bursa is separated from the distal half of the flexor pollicis longus sheath by a septum, making them separate sheaths.

The fifth finger flexor sheath commences at its terminal phalanx and, on reaching a point halfway up the palm, expands laterally to envelop the tendons of the 4th, the 3rd and the 2nd fingers. This expanded portion extends several centimeters proximal to the proximal volar crease of the wrist and is usually called the *ulnar bursa.*

Infrequently, the sheath of the index finger may extend to and communicate with the ulnar bursa. The 3rd and the 4th finger sheaths may also do this. Such variations much be kept in mind. Frequently, a communication exists between the radial and the ulnar bursae at the wrist. Infectious exudate may spread from one to the other, creating the so-called "horseshoe abscess."

Subtendinous Space (Parona's Space). The potential space between the superficially lying flexor digitorum profundus and the flexor pollicis longus tendons and the deeply lying pronator quadratus muscle is known as the *subtendinous space* or *Parona's space.* Infectious exudate can enter this space by rupturing the radial or ulnar bursa above, or by extension from a thenar or midpalmar space infection in the palm.

Lumbrical Muscle Sheaths (Plate 48). Each lumbrical muscle is enveloped in a thin transparent membrane. The sheath of the first lumbrical is in contact with and adherent to the membrane covering the thenar bursa. The sheaths of the 2nd, the 3rd and the 4th lumbricals overlie the midpalmar bursa. These sheaths extend from the web areas to the center of the palm. Thus, a web space infection can ascend a lumbrical canal and rupture into the thenar or midpalmar space; or infection in the thenar space or the midpalmar space can erode and descend a lumbrical canal to enter the web area.

Thenar and Midpalmar Spaces (Plate 48). The thenar space extends mediolaterally from the 3rd metacarpal bone to the thenar emi-

nence and proximodistally from the transverse carpal ligament to a line a thumb's breadth proximal to the webs of the fingers. The midpalmar space extends lateromedially from the 3rd metacarpal bone to the hypothenar eminence and proximodistally about 1 cm. more proximally than the thenar space. Each space is lined with a very thin membrane whose surfaces are in contact so that no space can be said to exist unless distended by an accumulation of exudate.

Unlike the flexor tendons of the thumb and the little finger, the flexor tendons of the index, the middle and the ring fingers have no sheath in their central palmar portions. The thenar bursa is interposed between the deep aspect of the index finger flexor tendon and its associated lumbrical and the superficial surface of the adductor pollicis. The thenar bursa extends about the distal border of the adductor to gain the dorsal aspect of the muscle and is interposed between the adductor and the 1st dorsal interosseous, the 2nd metacarpal and the 1st palmar interosseous. Similarly, the midpalmar bursa is interposed between the flexor tendons and the ensheathed lumbricals of the 3rd, the 4th and the 5th fingers, and the underlying metacarpals and interossei.

The proximal end of the index finger sheath and the sheath of the 1st lumbrical are in contact with the thenar bursa. Infection from the former may enter the latter. The proximal ends of the 3rd, the 4th and the 5th tendon sheaths and associated lumbrical sheaths are in contact with the midpalmar bursa. Suppuration of the former may extend to the latter.

RECONSTRUCTION OF THE HAND

Outline of Procedures

1. **Position of function maintained.** Wrist dorsiflexed, thumb opposing the fingers
2. **Order of operations**
   A. Remove cicatrix
   B. Provide good skin covering and subcutaneous fat
   C. Repair and immobilize the bone
   D. Mobilize joints
   E. Repair nerves
   F. Repair tendons
3. **Tourniquet.** Prolonged ischemia promotes tissue reaction; 1 hour is safe
4. **Anesthesia.** Block or general. Avoid local infiltration.
5. **The softening-up process.** Tissue induration follows trauma, infection and operation. Exercise and time itself, even a year, results in softening up of the tissues which are better nourished and tolerate surgical trauma better. Each consecutive step in operating must await the return to the well-nourished state. A 4-month waiting period is allowed between procedures.

Operative Technic

Preoperative soap and water cleansing and painting with antiseptic solution. The arm is draped on an arm-board. Nontouch technic is used.

Delicacy of tissue handling is essential. Incisions should not overlie or parallel tendons. Instead, they should be at a distance from structures repaired, following the skin flexion creases where possible. The exposure of the palm is made through an L-shaped incision which follows the distal crease, then turns proximally in the immobile heel of the palm to the center just beyond the wrist. This cuts through the palmar aponeurosis, and a combined skin-aponeurotic pad is lifted away from the underlying blood vessels and nerves. Dissection should start in normal tissue proximal or distal to scar tissue. Nerves should be dissected from above downward to preserve branches. Cicatrix is removed, and normal tissue is exposed. Tendons are grafted. Pulleys, ligamentous bands or tendon sheaths are cut through at one side and repaired later. Tendons are repaired first, and nerves at the end of the operation. Loose paratenon fat is removed from over the fascia lata or over the triceps muscle and interposed between tendon and any rough underlying bone or other tissue. A square section of this tissue is secured at each corner by No 000 catgut; strands are brought out through the skin and tied over a dressing. Then the hand is elevated, the tourniquet is released, and hemostasis is secured by compression and ligatures. The skin is closed. Postoperative hematoma leads to adhesions; therefore, tiny rubber drains are placed at operation, and a compression band-

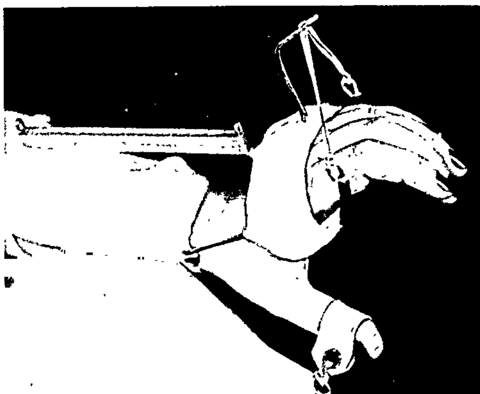




Fig. 323. Dynamic splinting of hand by use of plaster. The upper illustration demonstrates splinting for radial nerve paralysis. The lower shows correction in intrinsic muscle paralysis. The metacarpophalangeal joints are flexed, and the interphalangeal joints are extended. (Peacock, E. E : J. Bone & Joint Surg. 34A: 789)

age is applied. The drains are removed the following day.

Immobilization and elevation are necessary after operation After extensor tendon repair, the wrist and the fingers are maintained in dorsiflexion. The fingers themselves never should be immobilized in flexion to prevent flexor tendons from breaking because the tendons will adhere too far proximally and will limit extension. Merely flexing the wrist will deprive the flexor muscle of its breaking force. However, nerve repair may require flexion of various joints in order to approximate the nerve ends. Twenty-four hours later, bandages are loosened to allow for swelling. Drains are removed. Casts are bivalved.

Splinting

Plaster is applied on the dorsal and the volar aspects. Immobilization of a joint at the extreme of flexion should be avoided because the joint will be damaged. Position of function is sought, and thumb and finger are allowed free motion. The distal edge of the cast is at the distal crease on the palmar aspect and the knuckles on the dorsum. A volar slab maintains the wrist and the fingers in dorsiflexion after extensor tendon repair.

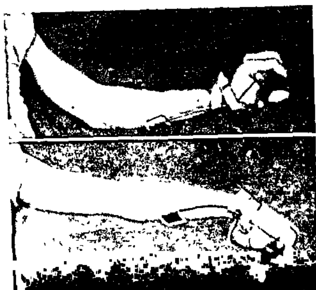


FIG. 324. Oppenheimer spring wire splint for radial palsy. (Dr. Sterling Bunnell)

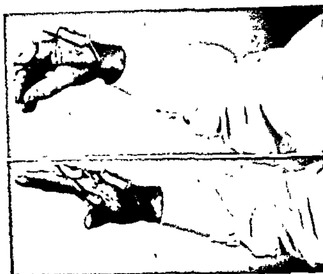


FIG. 325. Knuckle-bender splint to flex the metacarpophalangeal joints. (Dr. Sterling Bunnell)

A wire extension distally from a plaster cast may be used for traction. For avulsion of the distal insertion of the finger extensor tendon, reapproximation is attained by maintaining the distal joint in extension and the proximal in flexion by a plaster slab.

Special splints are used for immobilization, to maintain position, and to obtain flexion or extension gradually, to protect repaired tendons and nerves and to relax paralyzed muscles. Traction to flex the fingers should aim at causing the digits to converge toward a central point—the tubercle of the scaphoid. Prolonged splinting encourages stiffness. The basic splint is a wrist cockup splint, a volar forearm piece connected to a palm support and held to the hand and the forearm by encircling straps or laces. Various attachments and extensions may be added to attain the desired position of the fingers. A large rubber sponge in the palm of the hand affords compression. After plastic operations, the fingers are immobilized in a widespread position on a hand-spreading splint. This makes for ease of dressings. A glove with cords attached to the fingertips and looped through a buckle at the wrist can secure finger flexion effectively. Bunnell describes various types of splints. *The reader is referred to his classic* book on the hand. A dynamic splint can be constructed by use of plaster, rubber bands, and strong wire for hinges, lever arms and outriggers.

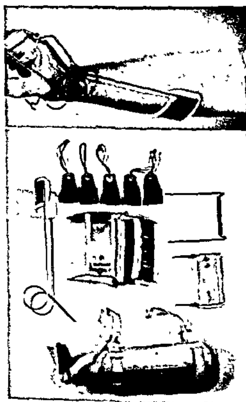


FIG. 326. (*Top*) Combination of spring Oppenheimer splint (to dorsiflex the wrist) and knuckle-bender splint (to flex the metacarpophalangeal joints) and attachment to extend the distal 2 finger joints. (*Bottom*) Parts and attachments for combination splint. (Dr. Sterling Bunnell)



Fig. 327. Tubular pedicle graft. The area between the thumb and the index finger is subjected to pressure requiring skin with good subcutaneous padding. The pedicle should be sufficiently long to permit relaxation during the period of transfer.

Treatment of Scars and Contractures

Open wounds and infections heal with extensive cicatrix which engulfs all structures and forms a relatively avascular area which does not heal readily after trauma or operative incision. Before attempting reconstruction of deep parts, the scar should be completely excised; the parts freed, and a covering of good skin with subcutaneous tissue placed in the defect. A sliding skin flap or pedicle graft is necessary. Free split-thickness grafts are inadequate.

Improper incisions will produce contractures. Median longitudinal incisions and those that cross skin creases at a right angle lead to keloids. In the finger a midlateral incision is proper. The palm is exposed by an incision which parallels the distal flexion crease and turns in an obtuse angle up the immobile heel of the palm to the carpus. A transverse incision at the wrist may be extended longitudinally upward and downward at either end At the pulp of the fingertip, a lateral or fishmouth incision is advisable. An incision which overlies a tendon becomes adherent to the

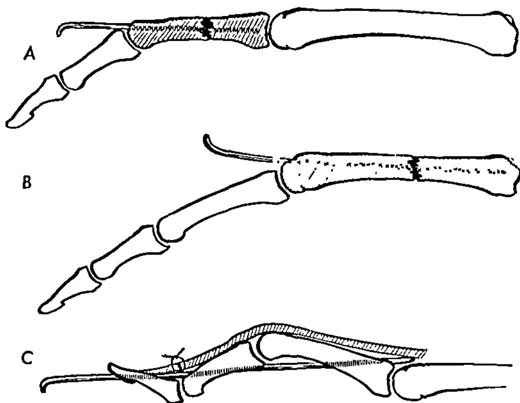


Fig. 328. Intramedullary pinning of fingers. (A) Fractured phalanx. (B) Fractured metacarpal. (C) Ruptured distal insertion of extensor aponeurosis.

tendon. Volar bands or pulleys should be opened to one side, not over the central gliding surface.

The removal of a keloid should be accompanied by a skin zigzag plastic or replaced with a graft which changes the direction of the skin incision.

Flexion contractures follow infections or burns. When treating infections, the position of function should be maintained on a splint. Infection in the palmar space paralyzes the intrinsic muscles, producing a claw hand. With subsequent cicatricial formation, the muscles are fibrotic, tendons become adherent, and the deformity is fixed.

Correction of Bony Deformities

Malunion of a fracture of the metacarpal or the phalanx results in abnormal rotation, angulation, and overriding of fragments. Rotation of the distal fragment allows the finger, during flexion, to cross over the adjacent finger. Angulation produces a dorsal or volar projection over which the tendon is stretched, thereby restricting its excursion. Tendons become adherent to the fracture site, especially in a phalanx. Treatment consists of open reduction of the fragments and inserting an intramedullary or inlay bone graft, to ensure union, because phalanges and metacarpals are slow to unite after surgical exposure. Postoperatively, a plaster splint is maintained for 5 to 8 weeks. Other fingers should be allowed freedom of movement. Loss of a metacarpal head of the long or the ring finger allows the remaining metacarpals to rotate so that their fingers during flexion cross each other. This is remedied by removing the remainder of the shortened metacarpal.

Intramedullary pin fixation[1] of fractured phalanges or metacarpals does away with plaster immobilization and allows continued movement of the hand and preservation of joint mobility. When a phalanx is fractured, the pin is inserted through the distal end of the phalanx while the joint at this level is held in 90° of flexion, and the pin crosses the fracture line and enters the proximal phalanx. The end of the wire protruding through the skin is bent to a right angle and covered with a cotton and collodion dressing. The wire is removed 4 to 7 weeks later, depending on union. Closed reduction is done in fresh fractures and open reduction in delayed or reconstructive procedures. Postoperatively, the patient is encouraged to move the finger wherein flexion is complete and extension about 150°. Full motion is recovered after the removal of the pin. The method is contraindicated in oblique fractures and in the presence of infections. A metacarpal fracture should be exposed surgically, the pin inserted at the fracture site through the distal fragment to emerge at the radial aspect of the dorsal surface of the metacarpal head while the metacarpophalangeal joint is maintained in full flexion. Then the pin is drilled into the proximal fragment to the base of the bone.

Stiffening of Joints

Synovial membrane, joint capsule and ligaments are supple and form folds when the joint is relaxed but are barely long enough to permit the extreme of motion when they are fully stretched out and hold the joint surfaces snugly together. Shortening of these structures, for example by serofibrinous exudate causing adhesions, will limit motion. The exudate which accumulates as a result of trauma contains fibrin. Fibrin, if allowed to persist, will be organized into fibrous tissue. When a joint is damaged and not immobilized immediately, it becomes swollen with edematous fluid. If a joint is splinted under strain, it is injured, becomes more edematous and eventually stiffens. Inactivity of the hand causes edema, because lack of muscle action permits the tissue fluids to stagnate.

Prevention of joint stiffening is accomplished by elevation, immobilization in midposition, and avoiding excessive manipulation. The uninjured parts must be kept moving.

Before tendons can be repaired, the affected joints must be mobilized. First, the position of function is obtained. The wrist is dorsiflexed, carpal and metacarpal arches are well curved, the thumb is placed in opposition, and metacarpophalangeal joints are flexed by mild continuous traction and use of buckle and elastic bands. The apparatus is removed at intervals to allow exercise of joints. Forcible manipulation is avoided, because it tears tissues and causes further scarring.

[1] vom Saal, F. H.: Intramedullary fixation in fractures of hand and fingers, J. Bone & Joint Surg. 35A:5-16, 87, 1953.

Surgery of Joints

Before operating on a joint, the presence of infection or rheumatic inflammation should be determined.

The wrist, following infection, usually becomes ankylosed in flexion, a nonfunctional position, and involvement of the lower radio-ulnar joint restricts rotatory forearm motion. Arthrodesis in dorsiflexion and removal of the lower end of the ulna corrects this. Old dislocation of the carpus may be reduced after excising the interposed obstructing soft tissue. Localized degeneration of the lunate or the navicular requires removal of the offending bone. If these conditions are of long-standing, a generalized degenerative arthritis of the wrist results and demands arthrodesis to eliminate disabling symptoms. Old non-union of the fractured navicular produces a similar situation. Removal of the proximal row of carpal bones has been done for painful stiffening radiocarpal arthritis but results in weakened grasp. Arthrodesis in dorsiflexion produces a strong, stable, painless wrist. Arthrodesis between 20° and 30° dorsiflexion and slight ulnar deviation produces the best finger balance. In paralytics, flexion of the fingers may be automatic, depending on active extension of the wrist. Therefore, arthrodesis should be avoided. Obliterating the wrist joint makes available 6 tendons for use in transplantation.

At the lower radio-ulnar joint, an articular disk of cartilage overlies the distal aspect of the ulna, is attached to the ulnar edge of the distal end of the radius, and is inserted into the styloid of the ulna. It constitutes a septum which extends between the synovial cavity of the carpal area distally and that of the radio-ulnar joint proximally. The 2 synovial enclo-

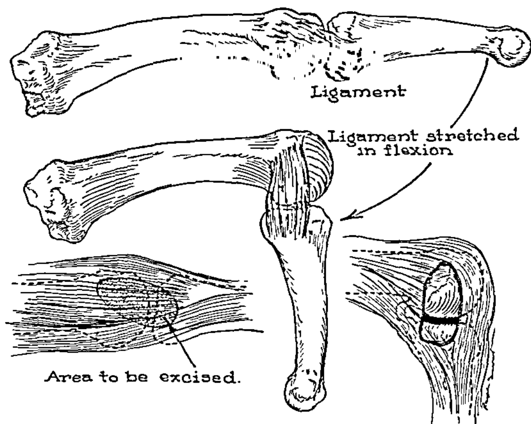


Fig. 329. Capsulectomy of the proximal finger joint. Normally, the collateral ligaments are slack in extension but tight in flexion. If these ligaments, because of edema and immobility, become short and thick, the joint will no longer flex Through a short longitudinal incision on each side of the knuckle, the joint capsule is exposed between tendons of the long extensor and the interosseus muscles and on excising the collateral ligament from each side the joint easily can be placed in flexion. (Bunnell, S.: Surgery of the Hand, ed. 3, Philadelphia, Lippincott)

sures frequently communicate and are affected by the same disease process. The styloid of the ulna is attached by a ligament to the triquetrum. This ligament is torn (or the styloid tip is avulsed) in flexion and extension injuries of the wrist. In Colles' fracture, the distal end of the ulna displaces anteriorly. In the reverse Colles' (Smith's) fracture in which the distal radial fragment is rotated anteriorly, the ulna displaces dorsally. Forced supination of the forearm will dislocate the ulnar head forward. A loose lower ulna will displace forward and backward with supination and pronation respectively, each movement being accompanied by a click due to the extensor carpi ulnaris slipping over the styloid. Pain and weakness of grasp result. Removal of the lower end of the ulna relieves pain due to a slipping tendon and radio-ulnar arthritis. It restores supination and pronation when these movements are hindered by a damaged or fixed radio-ulnar joint.

Collateral ligaments have a tendency to shorten and thicken and restrict joint motion. The metacarpophalangeal joints become fixed in extension. Excision of these ligaments restores motion. If the joint is destroyed, arthroplasty gives satisfactory function.

The interphalangeal collateral ligaments are stretched to their fullest extent at right-angle flexion. Therefore, contracture of the ligaments can occur with the finger flexed beyond this point, preventing extension. Capsulotomy and arthroplasty are ineffectual. Arthrodesis in moderate flexion is best from a functional standpoint.

Arthrodesis of the interphalangeal or the metacarpophalangeal joint of the thumb in moderate flexion gives an excellent functional result. The carpometacarpal joint, if stiffened with the thumb in abduction, requires an osteotomy through the metacarpal with placement of the thumb in the functional position of opposition. The dorsal lateral ligaments of the metacarpophalangeal joint ruptures from a blow, as in boxing, causing displacement of the base of the first phalanx and instability. Early treatment consists of immobilizing the thumb in full extension and abduction. Later, reconstruction of the ligaments is necessary. Dislocation at the metacarpophalangeal joint occurs by the mechanism of violent hyperextension. The metacarpal head penetrates between the 2 heads of the short flexor. The interposition of these tendons and the capsule creates an obstacle to reduction and necessitates surgical exposure. The carpometacarpal joint may be arthrodesed only if the navicular is not involved by disease and able to compensate for loss of motion. Otherwise, arthroplasty is indicated. Instability of the carpometacarpal joint results if the capsule is torn or if the volar edge of the metacarpal base which normally fits over the multangular bone is fractured and malunited. *The base of the metacarpal slips backward with pain and weakness each time an attempt is made to grasp.* Surgical correction by wedge osteotomy with the base dorsally and reinforcing the ligaments results in a stable thumb.

Repair of Nerves to the Hand

Repair of deep nerves is simple, and regeneration is excellent. Preservation of sensation is essential to good function. Severance of the tiny motor thenar nerves results in loss of power to oppose the thumb to the fingers. If the deep branch of the ulnar nerve is severed, a clawhand results, with weakness in grasp of the thumb and inability to spread the fingers. A finger deprived of its nerve becomes atrophic and does not heal well. Therefore, nerve repair and regeneration should precede other operations. Regeneration of a volar digital nerve takes place at about ¾ in. per month. The rate of regeneration is better when cicatrix is minimal and nutrition of tissues is good. Sensory perception returns in the following order: coarse touch, pain (pinprick), light touch (cotton, wool) and stereognosis. Tinel's sign demonstrates a tingling or tickling sensation over the area of newly acquired sensation. When sensation is restored, the trophic effect of the severed nerve disappears, and the condition of the finger improves. When motor branches to the intrinsics are repaired, return of muscle function requires more than a year.

Neuromata of the Hand

A severed nerve forms a proliferating coil of neuraxons and Schwann cells interlaced with the scar of repair. This neuroma may be the source of persistent pain. Typically, such pain is chronic, episodic and initiated by thermal, mechanical and psychic trauma. It

produces vasomotor and sudomotor phenomena. Pain radiates to other areas. Sympathetic block anesthesia will relieve it for a prolonged period. Neurectomy does not relieve pain for more than a short time. Removal of the neuroma and surrounding cicatrix is urgently indicated. Occasionally, small loosely adherent neuromata in the finger or palm may be freed by massage, percussion, or injection under pressure of a fluid, such as a procaine solution.

Pathologic Characteristics of Tendons

A tendon is surrounded by a gliding mechanism. When it pulls in a straight line it is surrounded by and attached to specialized, loose, elastic fatty tissue—the paratenon. *Examples are found on the dorsum of the* foot and on the distal forearm, volar and dorsal. In the palm, the flexors of the first 3 fingers are surrounded by paratenon. A tendon which glides around a curve or a bend is surrounded by a thin tendon sheath. This consists of 2 layers: a visceral one (epitenon), enveloping the tendon, and a parietal one (sheath), lining the inner aspect of a thick fascial tunnel. The parietal layer is connected to the visceral layer by a mesotenon which attaches to the tendon on its convex side and carries the blood supply to it. Where the tendons pull on the flexor aspect of fingers, they are prevented from bowstringing by strong fascial coverings, the annular ligaments which are located over the phalanges and not over the joints.

A tendon severed within its sheath makes no effort to proliferate and heal. The tendon ends become rounded over, and the proximal end retracts. *However, severance outside the sheath is followed by proliferation whereby the proximal segment reaches out in an effort to reattach and becomes adherent to surrounding structures.* The proliferation is more pronounced if associated infection is present. Infection under pressure within a firm sheath will cause necrosis and destruction of the tendon and scar tissue replacement The cicatrix is extensive and binds the finger into severe flexion contracture which cannot be overcome by nonsurgical methods. The amount of tendon retraction depends on the tendon amplitude. *Within the first 2 months, the tendon may be drawn down and repaired. After that time, the muscle contraction prevents this, and tendon grafting is necessary.* Tendons degenerate from disuse, and they become unfit for use as grafts. *When a tendon is adherent at one place, it is also adherent from that point distally.*

The proliferation of a tendon end, when severed in paratenon, may reach out as a prolongation thicker than the tendon itself, attaching to surrounding structures, and may even reach the distal segment in an attempt to re-establish continuity of the tendon. Adherence to surrounding structures not only restricts its own gliding but hinders the action of the common muscle on other fingers. A flexor profundus tendon adherent in the palm *will restrict flexion of the adjoining fingers* and limit action of its antagonist extensor. Freeing it in the palm is insufficient, because it will reattach. It should be removed completely and replaced by a free tendon graft extending above the wrist. A free end of tendon will always reattach to surrounding tissue. It should be imbedded in an adjoining tendon.

Direct trauma or infection may produce a hand swollen by serofibrinous exudate. Fibrous replacement will result in adhesions which in turn incarcerates the tendons. Prophylaxis is attained by continuous early motion, elevation and *avoiding prolonged splinting.*

In healing of tendons when 2 cut tendon ends are approximated, they become reddened and swollen and engulfed by a translucent substance into which capillaries and fibroblasts penetrate. This fibroblastic bulbous connection is soft and friable at the end of 1 week The ingrowth of granulomatous fibroblastic reaction occurs from the surrounding tissue and continues through the 2nd week, but actual formation of tendon collagen fibers does not occur until the 3rd week. The tendon juncture becomes firm, and cleavage occurs between the fusiform mass and the surrounding tissue prior to movement. *At the end of the 4th week, swelling and vascularity have decreased, and the tendon has loosened sufficiently for gliding motion. The junction is now strong.* Repair in paratenon formation is more rapid and is stronger than within a sheath. Before 3 weeks, excessive motion

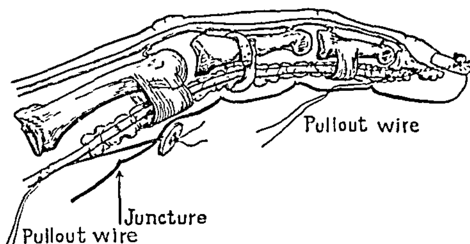


Fig. 330. Diagram of reconstruction of a tendon in a finger which is quite cicatricial. A free tendon graft plus its paratenon has been threaded through 3 pulleys (2 natural and 1 reconstructed). The graft is sutured to the profundus in the palm and to the distal phalanx where adhesions will do the least harm. The distal phalanx is first scraped for bony contact. Suturing is with removable stainless-steel wire, the proximal one being a "suture at a distance." (Bunnell, S.: Surgery of the Hand, ed. 3, Philadelphia, Lippincott)

causes reaction, proliferation, adhesions, and separation of tendon ends. The tendon should be immobilized during this time. After 3 weeks, tissue reaction is less in response to motion, and the junction becomes stronger. Guarded restricted exercises during the 4th and the 5th weeks may be followed by strong unlimited motion which will strengthen and mobilize the tendon.

A tendon must glide freely. It will become adherent in a scarred area and in poorly nourished tissues. The scar should be excised and the surrounding skin undermined and allowed to retract for better nutrition, and the open defect should be replaced by a pedicle graft, containing adequate subcutaneous fat. To prevent adherence, the tendon is surrounded by slippery material, either paratenon or smooth deep fascia. Paratenon, a layer of specialized fat, is obtained from over the triceps aponeurosis or from over the deep fascia or the fascia lata in the lower thigh, particularly its posterior aspect. It directly overlies the deep fascia. Thin deep fascia, the undersurface of which is slippery, is obtained from the front or the back of the forearm or over the anterior thigh. A patch of tissue is placed between the tendon and the cicatricial bed, and its edges are kept at a distance medially and laterally by catgut sutures. This method is used to free tendons which have become adherent after repair or over the site of a fracture.

When a tendon graft is withdrawn, a thin slippery gliding layer of tendon sheath adheres to the surface and provides insulation against adhesions. For better covering, a tendon graft may be removed with its surrounding paratenon and transferred as a complete gliding unit. The palmaris longus or the long extensors of the toes may be used. An alternative is to place a sheet of paratenon about the tendon at the time of grafting or later at a tendon-freeing operation.

The site of tendon suture should be selected with care. The junction will adhere in a firm fascial tunnel as in the wrist and the fingers. In the distal palm it adheres to the annular sheath over the metacarpal head or to the fascial septa. *Suturing is done in the proximal part of the palm or in the forearm* where the tissues are soft and movable.

Tendon Grafts

After a tendon is severed and several months have elapsed, it degenerates about the severed ends and is unsuitable for repair. The proximal segment cannot be pulled distally. A tendon graft is advisable. During the early weeks the tendon graft is very swollen and surrounded by vascular tissue. *Repair is slower than in normal tendon, so that strength at the suture line is attained at about 5 weeks.* In healing there is a tendency to contract; consequently, the tendon graft should be a little longer than the gap to be bridged. The palmaris longus is an excellent source for a

graft. Its presence is demonstrated by flexing the wrist against resistance. A small transverse cut is made at each end, and the tendon is withdrawn distally. It will be found surrounded by a slippery epitenon. If additional slippery material is required, a longitudinal incision is made over the tendon, and the latter is removed with the surrounding epitenon fat. *The flexor sublimis and the profundus tendons should not be repaired in the same finger, because they will adhere to each other.* Therefore, through a lateral incision over the middle phalanx, the insertions of the sublimis are severed and withdrawn at the wrist and may be used to bridge a gap in the profundus. The long extensors of the toes are obtained by severing them at the base of the toes, sparing the brevis tendons. Through a high short transverse incision on the dorsum of the foot, the tendons are freed from paratenon by running a tendon stripper distally and then withdrawing the tendons. Then another incision is made longitudinally lateral to the anterior tibial. The tendons are pulled upward after stripping off the tendon of the peroneus tertius.

Tendon Transfers

The tendon of a functioning muscle can be substituted for a paralyzed muscle by transferring it to the bone or the tendon of the latter muscle The following principles should be observed:

1. Joints proximal to parts to be moved should be stabilized, either by tendon action or by arthrodesis.

   A. *Thumb pinch action.* Stabilize carpometacarpal joint in extension and metacarpophalangeal joint in flexion.

   B. *Finger extension* Metacarpophalangeal joint is maintained in slight flexion; at least, it is prevented from hyperextending.

2. Correct deformity first by shaping bones and joints.
3. Muscle of transferred tendon should be healthy.
4. Similar function in transferred muscle is desirable.
5. Gliding bed
6. Pull in straight line
7. Amplitude of motion is sufficient Do not attach tendon to multiple tendons of different amplitude.

The wrist can be arthrodesed in dorsiflexion, making all wrist tendons available for transfer. When paralysis is extreme, finger tendons may be tenodesed in the forearm so that flexion or extension of the wrist will produce automatic movements in the fingers.

Transfers for Radial Nerve Paralysis. Loss of extension of the wrist, the thumb and the proximal phalanges must be overcome, and abduction of the thumb provided. Three tendons are needed. One is attached to the long extensors of the long, the ring and the little fingers; the second, to the long extensor of the index finger and the thumb, because these usually work together; the third, to the wrist dorsiflexors and to the long abductor of the thumb, which actions occur simultaneously. Tendons utilized are the flexor carpi ulnaris, the flexor carpi radialis, the pronator teres, and/or the palmaris longus. When the radial nerve is involved at the level of the posterior interosseous nerve, the wrist dorsiflexors are spared, and the extensor carpi radialis brevis is used to extend the fingers and the thumb; the palmaris longus is used to abduct the thumb.

Median Nerve Paralysis. Flexion of the index and the middle fingers, flexion and opposition of the thumb, and radial flexion of the wrist are lost. The upper end of the profundus tendons of the index and the middle fingers can be attached to those of the ring and the little fingers and reinforced by a supinator longus transfer. The extensor carpi radialis brevis is attached to the flexor pollicis longus, and the flexor carpi ulnaris tendon can be routed through a pulley at the pisiform bone to furnish opposition to the thumb.

Ulnar Nerve Paralysis. If the profundus to the little and the ring fingers is involved, partial paralysis results, because the median nerve provides some innervation Tendon transfer is required only for loss of action of intrinsic muscles (to be described later).

Median and Ulnar Nerve Paralysis. *Arthrodese the wrist in 30° dorsiflexion.* Then the wrist extensors are available for transfer to the profundi of the fingers and the long flexor

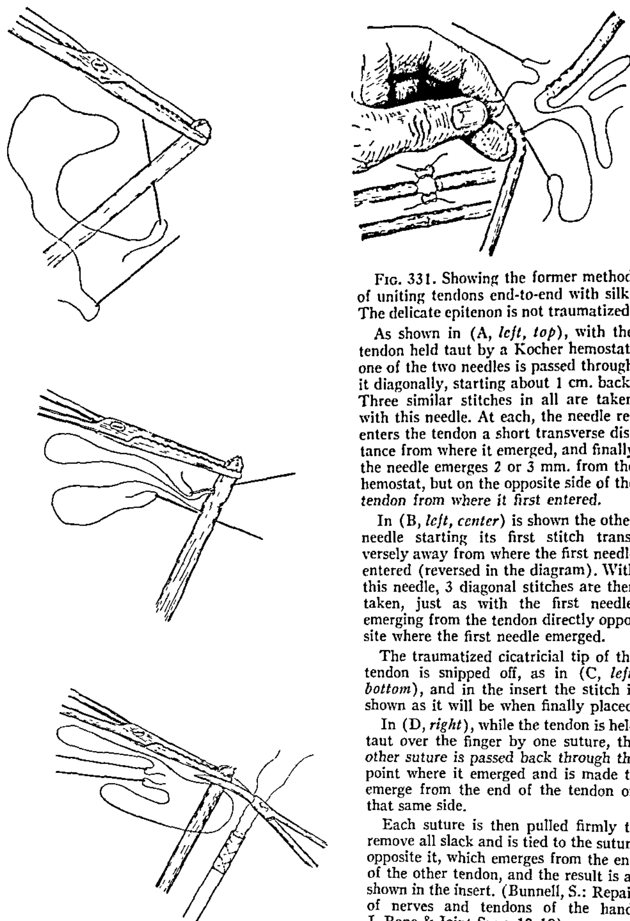


FIG. 331. Showing the former method of uniting tendons end-to-end with silk. The delicate epitenon is not traumatized.

As shown in (A, *left, top*), with the tendon held taut by a Kocher hemostat, one of the two needles is passed through it diagonally, starting about 1 cm. back. Three similar stitches in all are taken with this needle. At each, the needle re-enters the tendon a short transverse distance from where it emerged, and finally the needle emerges 2 or 3 mm. from the hemostat, but on the opposite side of the *tendon from where it first entered.*

In (B, *left, center*) is shown the other needle starting its first stitch transversely away from where the first needle entered (reversed in the diagram). With this needle, 3 diagonal stitches are then taken, just as with the first needle, emerging from the tendon directly opposite where the first needle emerged.

The traumatized cicatricial tip of the tendon is snipped off, as in (C, *left, bottom*), and in the insert the stitch is shown as it will be when finally placed.

In (D, *right*), while the tendon is held taut over the finger by one suture, the *other suture is passed back through the* point where it emerged and is made to emerge from the end of the tendon on that same side.

Each suture is then pulled firmly to remove all slack and is tied to the suture opposite it, which emerges from the end of the other tendon, and the result is as shown in the insert. (Bunnell, S.: Repair of nerves and tendons of the hand, J. Bone & Joint Surg. 10:10)

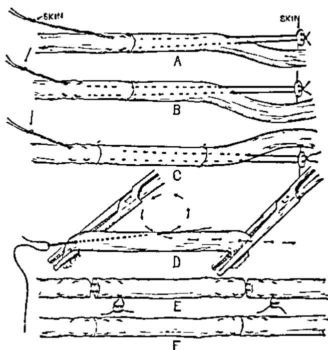


FIG. 332. Methods of tendon suture. (A, B, C) Stainless-steel pull-out sutures. (A) Threading the suture through the distal tendon end holds the tendon ends in good approximation. The pull-out wire is twisted to prevent tissue growing between and hindering withdrawal. (B) Placing the splicing part of the suture at a distance to lessen adhesions at the important place, the junction. (C) A short graft may be threaded on the suture wires A tiny stitch of the finest silk keeps each end of the graft opposed to the tendon end. (D) Method of threading the sutures longitudinally through a tendon. (E, F) Inserting a graft between tendon ends using a silk suture. Short grafts may be threaded on the silk as in (C). (Bunnell, S : Surgery of the Hand, ed 3, Philadelphia, Lippincott)

of the thumb The extensor carpi ulnaris can be prolonged by a sublimis graft to furnish thumb opposition. When arthrodesis and tendon transference is not desirable, tenodesis of the flexor profundus and the flexor pollicis longus tendons above the wrist will provide automatic finger flexion as the wrist is actively extended. The thumb may be fixed in permanent opposition by a bone graft between the first 2 metacarpals (Thompson operation).

Radial and Ulnar Nerve Paralysis. The wrist should be arthrodesed in dorsiflexion. The flexor carpi radialis is transferred to the long extensors of thumb and the index finger. The pronator teres is used to extend the long, the ring and the little fingers. The palmaris longus is transferred to the long thumb abductor. The flexor sublimis is used to restore intrinsic muscle action.

Pronation Deformity. The flexor carpi ulnaris and the pronator teres are routed about the ulnar side to the dorsal aspect of the forearm and inserted into the distal radius The flexor carpi ulnaris may be substituted. In addition, the pronator quadratus may be severed, and a rotation osteotomy of the ulna is done.

Technic of Tendon Repair

All scar tissue is removed by sharp knife dissection down to normal tissue. A tendon stripper is applied at the proximal normal tendon and by twirling motion works its way distally around the surface of the tendon. When freeing a tendon within a finger, an incision is made mid-laterally along the entire length of the finger, and knife and stripper are used under direct vision. A rough and adherent tendon should be excised, because it will only re-adhere. Reforming of adhesions is prevented by sharp knife dissection, delicate handling, hemostasis, rubber tube drains, compression dressings, splinting, postoperative elevation and early motion. When a tendon must lie on a cicatricial bed or on raw bone, interposition of gliding material is necessary. When suturing tendons end to end, it is necessary to keep the suture material within the inner part of the tendon and to remove the tension from the tendon ends.

Silk Technic. Untreated silk (.012 in) is used because treated silk slips at the knot. It is threaded at each end on a straight spear-pointed needle. The tendon end is grasped by a hemostat and drawn taut. The needle is started from ½ to ¾ in. from the end and is passed transversely across the tendon. Then each needle is passed obliquely back and forth 3 times, always re-entering the tendon a short distance from where it started. The traumatized end of the tendon is cut off, and then each needle is passed distally through the tendon to emerge from the raw cut surface. Each suture is pulled taut, and all slack in the tendon is taken up. Then the threads are tied to

2 threads similarly placed in the distal segment of the tendon.

An alternative method is to continue the original 2 needles into the distal segment in reverse fashion, both threads emerging at the same point and there tied. When bridging a gap, a free tendon graft is interposed between the 2 segment ends, the needles are passed straight through the graft, and the suture in the distal segment is completed. A fine approximation suture of blood vessel silk is inserted at both junctures.

Removable Stainless Steel Wire Technic. Stainless steel wire causes no foreign body reaction and is strong. It will break when placed over joints where it is subject to constant angulation movement. Therefore, it should be used where the tendons pull in a straight line. It is spliced in the proximal segment only, which it holds distally and thereby removes tension from the line of junction. After the tendon has united at 3 weeks, the wire is removed, thereby eliminating mechanical irritation. Wires used are gauge 30 for very large tendons, 34 for hand flexor tendons, and 35 for small tendons. In placing steel wires, great caution is necessary to avoid kinking, because the wire may break. The following are the different methods used:

1. END-TO-END. Wire is passed through the proximal tendon in crisscross fashion. At the proximal loop of this wire, a second wire is looped through it and twisted on itself and passed proximally in the line of direction of the tendon and emerges through the skin. This latter is the pull-out wire. The 2 wires which emerge from the cut end of the tendon are passed distally uncrossed through the distal segment for about 1 inch, then diagonally out through the skin where they are tied over a button while tension is exerted, drawing the proximal tendon distally.

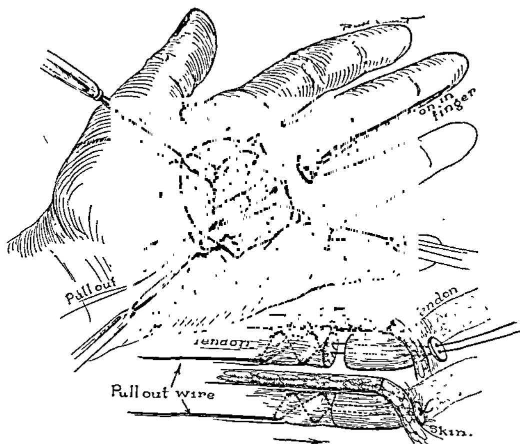


FIG. 333. Suturing a tendon in the palm by removable stainless-steel wire. In 3 weeks, the tendon will have joined and the suture after being cut beneath the button may be withdrawn backward by the pull-out wire, which is looped about it. (Bunnell, S.: Surgery of the Hand, ed. 3, Philadelphia, Lippincott)

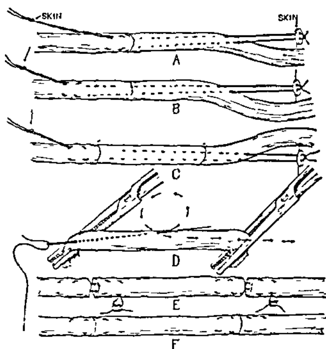


FIG. 332 Methods of tendon suture. (A, B, C) Stainless-steel pull-out sutures. (A) Threading the suture through the distal tendon end holds the tendon ends in good approximation The pull-out wire is twisted to prevent tissue growing between and hindering withdrawal. (B) Placing the splicing part of the suture at a distance to lessen adhesions at the important place, the junction. (C) A short graft may be threaded on the suture wires. A tiny stitch of the finest silk keeps each end of the graft opposed to the tendon end. (D) Method of threading the sutures longitudinally through a tendon (E, F) Inserting a graft between tendon ends using a silk suture. Short grafts may be threaded on the silk as in (C). (Bunnell, S.: Surgery of the Hand, ed. 3, Philadelphia, Lippincott)

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and the profundus tendons. In the distal and the proximal parts of the palm, sheath formation is present. *Tendon junctures in the distal palm are apt to become adherent.* A tendon juncture made in the region of the annular sheath or pulley opposite the metacarpal head will become firmly adherent. *A tendon graft should be placed from the base of the palm to the end of the finger.*

Within the finger a tendon juncture usually adheres to the phalanx. *Suture over the proximal phalanx is contraindicated.* A primary suture occasionally succeeds at the level of the middle phalanx, but secondary suture should not be done. The sublimis is severed at its slips of insertion and withdrawn through a transverse incision above the wrist and used to bridge the gap in the profundus. When both tendons are severed in the same finger, the sublimis should be removed; otherwise, the tendons will adhere to each other, and motion will be greatly restricted. Pulleys, particularly those over the proximal phalanges, should be resutured or reconstructed. Before any reconstructive surgery is attempted, all scar is removed, and a good skin covering is provided. Nerves are repaired at the same time as the tendon. A mid-lateral incision is used. If primary suture is attempted within the finger, adhesions invariably form and restrict motion. Encircling the tendon with gliding tissue will correct this. If the sublimis is intact, flexion of the middle phalanx is preserved, and the profundus may be ignored.

Extensor tendon suture usually succeeds, because these tendons move freely in the forearm and the hand. When an extensor tendon is severed, a dropped finger at the metacarpophalangeal joint results. Sometimes no loss of function results, because the extensor tendons interdigitate in the hand, and an adjacent tendon will continue extensor motion. In the absence of sufficient tendon for suture, the extensor proprius of the index or the little finger may be transferred. The stub of the proprius should be attached to the remaining tendon of its finger so that the latter will not extend the finger in a rotatory manner.

Postoperative Treatment

After repair of flexor tendons, the fingers are allowed freedom of motion to avoid adherence. For extensor tendons, however, the wrist and the fingers are splinted in extension. Tiny rubber tube drains, pressure dressings, splinting and elevation reduce hematoma formation and fibrinous exudate which might encourage adhesions. Twenty-four hours later, the drains are removed, pressure is released, and splinting is continued. During the first 3 weeks, finger movement is continued but held to a minimum because free tendon grafts are acquiring a new blood supply. Excessive motion will encourage scar tissue formation and adhesions about the suture line. After 3 weeks, the wire sutures are removed, and exercises are instituted, gently at first, the splint being removed for exercises and replaced afterward. After the 4th week, exercises are unrestricted. When moving a flexor tendon, the proximal phalanx is held extended while the muscle contracts and flexes the proximal interphalangeal joint. Next, the proximal and the middle phalanges are held extended while the muscle flexes the distal interphalangeal joint. At the beginning, passive flexion of the joints is done while active flexion is attempted. By extending the finger passively, the flexor tendon moves distally. When adherence of the tendon in the palm is suspected, strong active flexion of the finger against resistance will cause the tendon to elevate from its bed, and adhesions may be freed. Violent manipulations must be avoided, because adhesions may be torn with production of hemorrhage and more adhesions. Exercises are continued for many weeks.

After tendon repair, during the early weeks, there is some induration about the tendon and limitation of motion. The tissues slowly soften over many months, and the tendon gradually increases its amplitude of motion. If restriction of motion persists after many months, adherence of the tendon to surrounding tissues may be assumed. The tendon should be exposed, freed and encircled with gliding tissue, and energetic motion should be instituted early. *Adherence of a tendon in the palm implies that the tendon distally from that point extending into the finger is likewise adherent and must be freed.*

THUMB TENDONS

Severance of the *extensor pollicis longus* results in loss of extension of the distal

2. End-to-End Suture at a Distance. This is done to keep the suture away from the tendon juncture, thereby minimizing tissue reaction at this site. The wire is placed in the proximal segment but emerges at a distance from the tendon end; then it is passed within the tendon sheath distally and out through the skin. A small suture of fine blood vessel silk approximates the tendon ends. This method is used in the finger where the tendon glides through narrow confines. A pull-out stitch is added.

Attaching Tendon to Bone. A crisscross wire suture with pull-out wire attached is applied to the tendon end. A flap of bone is elevated, and a hole is drilled through the bone. The wires are passed through the hole to the opposite surface and are tied over a button as the hand is flexed and the wires pulled taut.

Attaching Tendon to Distal Phalanx. The volar aspect of the base of the phalanx is scraped, and a hole is drilled through, the bit emerging through the nail. Wires are passed through the hole and tied on the nail surface.

Construction of Pulleys. Absence of a pulley will permit a tendon to bowstring and lose its efficiency. The pulley must be restored or reconstructed. A transverse carpal or dorsal carpal ligament is constructed by placing a band of fascia lata across the wrist and fixing it to bone at the side of the wrist.

Over the flexor aspect of the fingers, a proximal pulley extends from the level of the metacarpal head to the middle of the proximal phalanx. The distal one lies opposite the middle phalanx. To resuture a pulley which has been severed laterally at operation, a wire suture enters the skin dorsal to the incision, penetrates the periosteum, catches the volar edge of the pulley by a mattress stitch and finally retraces its path to the dorsum, where the ends are pulled taut and tied. The sheath is thereby drawn to the side of the phalanx between the tendon and the nerves.

If a flexor pulley has been destroyed, it can be reconstructed by passing a free tendon graft about the tendon and the phalanx, beneath the vessels and the nerves, and suturing it to itself at the side of the finger.

Tension for Sutured Tendons. Within the first 2 months, the tendon ends can be approximated under normal tension. Beyond this time, the muscle will have contracted too much to allow approximation, and a free tendon graft or tendon transfer will be necessary. When using a free graft, one must allow for subsequent shrinkage of the graft which would cause, in the case of a flexor tendon, a flexion deformity of the finger. When placing a graft, the wrist and the fingers should be positioned straight out and suturing done under slight tension. (See section on muscle tension under Muscle Physiology.)

Splinting After Tendon Suture. A flexor tendon repaired anywhere in its course requires splinting of the wrist in flexion, the cast extending only as far as the metacarpophalangeal joints and permitting a little movement of the fingers. For extensor tendons, the cast extends to the distal phalanx, the wrist is dorsiflexed, and the fingers are extended.

Surgical Principles

In the flexor aspect of the forearm, the sublimis with its individual pointed muscle belly units extends farther distally than the deeper profundus tendons and can easily be withdrawn upward from the palm. On the other hand, the profundus tendons distally *interdigitate* and therefore cannot be withdrawn from the palm by pulling on the proximal ends. Instead, the distal ends may be drawn upward from the palm into the forearm. When repairing a flexor tendon in the forearm, a pull exerted on the distal segment may not flex the finger because of light ad hesions which formed during the period o inactivity. These are freed by flexing the fir ger completely and passively and then pullin on the tendon.

In exposing the palm surgically, the incisi parallels the distal crease and turns proxim ward to the center of the heel of the pal penetrating the palmar fascia. The nerves the vessels lie superficial to the tendons are spared. Any nerves found severed are paired immediately. Sublimis tendons smaller and superficial to the profundus latter give attachment to the lumbr In the central area of the palm, the se enclosing the soft thin paratenon tis split through laterally, exposing the su

and the profundus tendons. In the distal and the proximal parts of the palm, sheath formation is present. *Tendon junctures in the distal palm are apt to become adherent.* A tendon juncture made in the region of the annular sheath or pulley opposite the metacarpal head will become firmly adherent. *A tendon graft should be placed from the base of the palm to the end of the finger.*

Within the finger a tendon juncture usually adheres to the phalanx. *Suture over the proximal phalanx is contraindicated.* A primary suture occasionally succeeds at the level of the middle phalanx, but secondary suture should not be done. The sublimis is severed at its slips of insertion and withdrawn through a transverse incision above the wrist and used to bridge the gap in the profundus. When both tendons are severed in the same finger, the sublimis should be removed; otherwise, the tendons will adhere to each other, and motion will be greatly restricted. Pulleys, particularly those over the proximal phalanges, should be resutured or reconstructed. Before any reconstructive surgery is attempted, all scar is removed, and a good skin covering is provided. Nerves are repaired at the same time as the tendon. A mid-lateral incision is used. If primary suture is attempted within the finger, adhesions invariably form and restrict motion. Encircling the tendon with gliding tissue will correct this. If the sublimis is intact, flexion of the middle phalanx is preserved, and the profundus may be ignored.

Extensor tendon suture usually succeeds, because these tendons move freely in the forearm and the hand. When an extensor tendon is severed, a dropped finger at the metacarpophalangeal joint results. Sometimes no loss of function results, because the extensor tendons interdigitate in the hand, and an adjacent tendon will continue extensor motion. In the absence of sufficient tendon for suture, the extensor proprius of the index or the little finger may be transferred. The stub of the proprius should be attached to the remaining tendon of its finger so that the latter will not extend the finger in a rotatory manner.

Postoperative Treatment

After repair of flexor tendons, the fingers are allowed freedom of motion to avoid adherence. For extensor tendons, however, the wrist and the fingers are splinted in extension. Tiny rubber tube drains, pressure dressings, splinting and elevation reduce hematoma formation and fibrinous exudate which might encourage adhesions. Twenty-four hours later, the drains are removed, pressure is released, and splinting is continued. During the first 3 weeks, finger movement is continued but held to a minimum because free tendon grafts are acquiring a new blood supply. Excessive motion will encourage scar tissue formation and adhesions about the suture line. After 3 weeks, the wire sutures are removed, and exercises are instituted, gently at first, the splint being removed for exercises and replaced afterward. After the 4th week, exercises are unrestricted. When moving a flexor tendon, the proximal phalanx is held extended while the muscle contracts and flexes the proximal interphalangeal joint. Next, the proximal and the middle phalanges are held extended while the muscle flexes the distal interphalangeal joint. At the beginning, passive flexion of the joints is done while active flexion is attempted. By extending the finger passively, the flexor tendon moves distally. When adherence of the tendon in the palm is suspected, strong active flexion of the finger against resistance will cause the tendon to elevate from its bed, and adhesions may be freed. Violent manipulations must be avoided, because adhesions may be torn with production of hemorrhage and more adhesions. Exercises are continued for many weeks.

After tendon repair, during the early weeks, there is some induration about the tendon and limitation of motion. The tissues slowly soften over many months, and the tendon gradually increases its amplitude of motion. If restriction of motion persists after many months, adherence of the tendon to surrounding tissues may be assumed. The tendon should be exposed, freed and encircled with gliding tissue, and energetic motion should be instituted early. *Adherence of a tendon in the palm implies that the tendon distally from that point extending into the finger is likewise adherent and must be freed.*

THUMB TENDONS

Severance of the *extensor pollicis longus* results in loss of extension of the distal

2. End-to-End Suture at a Distance. This is done to keep the suture away from the tendon juncture, thereby minimizing tissue reaction at this site. The wire is placed in the proximal segment but emerges at a distance from the tendon end; then it is passed within the tendon sheath distally and out through the skin. A small suture of fine blood vessel silk approximates the tendon ends. This method is used in the finger where the tendon glides through narrow confines. A pull-out stitch is added.

Attaching Tendon to Bone. A crisscross wire suture with pull-out wire attached is applied to the tendon end. A flap of bone is elevated, and a hole is drilled through the bone The wires are passed through the hole to the opposite surface and are tied over a button as the hand is flexed and the wires pulled taut.

Attaching Tendon to Distal Phalanx. The volar aspect of the base of the phalanx is scraped, and a hole is drilled through, the bit emerging through the nail. Wires are passed through the hole and tied on the nail surface.

Construction of Pulleys. Absence of a pulley will permit a tendon to bowstring and lose its efficiency. The pulley must be restored or reconstructed. A transverse carpal or dorsal carpal ligament is constructed by placing a band of fascia lata across the wrist and fixing it to bone at the side of the wrist.

Over the flexor aspect of the fingers, a proximal pulley extends from the level of the metacarpal head to the middle of the proximal phalanx. The distal one lies opposite the middle phalanx To resuture a pulley which has been severed laterally at operation, a wire suture enters the skin dorsal to the incision, penetrates the periosteum, catches the volar edge of the pulley by a mattress stitch and finally retraces its path to the dorsum, where the ends are pulled taut and tied The sheath is thereby drawn to the side of the phalanx between the tendon and the nerves

If a flexor pulley has been destroyed, it can be reconstructed by passing a free tendon graft about the tendon and the phalanx, beneath the vessels and the nerves, and suturing it to itself at the side of the finger.

Tension for Sutured Tendons. Within the first 2 months, the tendon ends can be approximated under normal tension. Beyond this time, the muscle will have contracted too much to allow approximation, and a free tendon graft or tendon transfer will be necessary. When using a free graft, one must allow for subsequent shrinkage of the graft which would cause, in the case of a flexor tendon, a flexion deformity of the finger. When placing a graft, the wrist and the fingers should be positioned straight out and suturing done under slight tension. (See section on muscle tension under Muscle Physiology.)

Splinting After Tendon Suture. A flexor tendon repaired anywhere in its course requires splinting of the wrist in flexion, the cast extending only as far as the metacarpophalangeal joints and permitting a little movement of the fingers. For extensor tendons, the cast extends to the distal phalanx, the wrist is dorsiflexed, and the fingers are extended

Surgical Principles

In the flexor aspect of the forearm, the sublimis with its individual pointed muscle belly units extends farther distally than the deeper profundus tendons and can easily be withdrawn upward from the palm. On the other hand, the profundus tendons distally interdigitate and therefore cannot be withdrawn from the palm by pulling on the proximal ends. Instead, the distal ends may be drawn upward from the palm into the forearm. When repairing a flexor tendon in the forearm, a pull exerted on the distal segment may not flex the finger because of light adhesions which formed during the period of inactivity. These are freed by flexing the finger completely and passively and then pulling on the tendon.

In exposing the palm surgically, the incision parallels the distal crease and turns proximalward to the center of the heel of the palm, penetrating the palmar fascia. The nerves and the vessels lie superficial to the tendons and are spared Any nerves found severed are repaired immediately. Sublimis tendons are smaller and superficial to the profundus; the latter give attachment to the lumbricals. In the central area of the palm, the septum enclosing the soft thin paratenon tissue is split through laterally, exposing the sublimis

THE INTRINSIC MUSCLES

The tendons of the interossei and the lumbricals insert into the sides of the dorsal aponeurosis and give off transverse fibers which form a sling over the proximal phalanx. They form lateral bands which continue distally on each side of the finger to insert into the base of the distal phalanx. The lateral bands send fibers to insert on and extend the middle phalanx. At the base of the proximal phalanx, a slip from the deep surface of the long extensor tendon extends the phalanx and proceeds distally to insert between the two lateral bands upon the base of the middle phalanx. A small fibrocartilaginous disk on the undersurface of the dorsal aponeurosis overlies the metacarpophalangeal and the interphalangeal joints.

The dorsal hood or sling is important in flexion and extension of the proximal phalanx. When the long flexors bend the distal 2 phalanges, the long extensor relaxes, and the hood shifts distally, coming to lie over the shaft of the proximal phalanx which can now be pulled volarward by the intrinsics. The hood is shifted back over the metacarpophalangeal joint by pull of the long extensor, and leverage upon the phalanx by the hood is lost. However, pull of the intrinsic is transferred to the lateral bands, thereby extending the distal 2 phalanges.

At the proximal interphalangeal joint, the lateral bands send fibers medially to insert on and extend the base of the middle phalanx. During finger flexion, these lateral bands are displaced volarward by a lateral attachment to the collateral ligament on each side of the joint. This volar shift enables the band to gain several millimeters in a more direct route to the distal phalanx, enabling the latter to be flexed. By maintaining the proximal interphalangeal joint in flexion, the aponeurosis may be approximated more easily to the distal phalanx during repair.

The *lumbricals* are 4 small muscles, each of which arises from a profundus tendon, passes radialward and distally in front of the transverse metacarpal ligament (behind which lies the interosseous), then turns dorsally to join the interosseous tendon in forming the dorsal hood and the lateral band. The lumbrical extends the distal 2 joints, flexes the proximal phalanx and produces radial lateral motion to the finger. When the proximal phalanx is held stabilized in full extension by the long extensor, lumbrical extension of the distal 2 joints is strong. As the proximal phalanx is gradually flexed, the lumbrical extensor action becomes proportionately less, until at 45° it is nil. Beyond 45° the long extensor alone acts to extend the distal 2 phalanges. The lumbrical acts on the proximal phalanx during the entire range of flexion.

The 4 dorsal *interossei* abduct the fingers from the long finger as an axis. The 3 volar interossei adduct the fingers toward the axis. Each inserts into the lateral tubercle at the base of the proximal phalanx and also contributes lateral fibers in forming the dorsal hood before continuing distally in the lateral band. Thus the interosseous, like the lumbrical, can flex the proximal phalanx. When the proximal phalanx is stabilized in full extension, it extends the distal phalanges and imparts lateral motion to the fingers. The exception is the first dorsal interosseous which, with the long extensor taut, strongly abducts the index finger and aids pinching between the index finger and the thumb. The interossei with the lumbricals add strength to grasp; in ulnar paralysis, the grip is two thirds normal. As the proximal phalanx is flexed increasingly, the effect on extending the distal phalanges is gradually diminished and progressively taken over by the long extensor. Beyond 45°, the long extensor alone is operative in extension. All interossei are innervated by the ulnar nerve.

The long extensor tendon extends the proximal phalanx and acts to extend the distal 2 phalanges in synergism with the intrinsics. During the first 45° of flexion of the proximal phalanx, the power of the extension by the long extensor increases and by the intrinsics decreases until, at 45° and beyond, the long extensor acts alone. During the last quarter of flexion, the long extensor is helpless to act on the distal phalanx, because the attachment to the base of the proximal and the middle phalanges limits its excursion. The long extensor also stabilizes the proximal phalanx in extension, so that intrinsic muscles can extend the distal phalanges and impart lateral motion to the finger.

phalanx and weakening of extension of the proximal phalanx, the brevis continuing its function alone. When the longus is adherent at the wrist, it will limit the motion of opposition. Where the tendon passes through its groove on the dorsum of the radius, it is subject to friction, particularly when the bony surface becomes irregular after a Colles' fracture. Therefore, excessive use of the thumb predisposes to spontaneous rupture (drummer's palsy). It may be repaired by suture or tendon graft, or an extensor of the index finger may be transferred.

The *extensor pollicis brevis* is important only for its use in tendon transfer in restoring opposition to the thumb.

The *abductor pollicis longus*, which inserts on the base of the 1st metacarpal, stabilizes the latter in extension in the act of pinching. Without it, the metacarpal rides forward, the metacarpophalangeal joint becomes hyperextended, and pinch is ineffectual.

De Quervain's Disease

The tendons of the abductor longus and the extensor brevis at the styloid of the radius pass through a thick tendon sheath which frequently becomes stenosed and inflamed and produces symptoms. This condition usually occurs in occupations requiring frequent thumb pinching and wrist movement. Pain develops about the radial styloid, radiating distally along the thumb and proximally into the forearm and is aggravated by thumb movement. The pain is increased when the tendon is angulated at the wrist by dorsiflexion, volar flexion, or abduction of the hand at the wrist. When the thumb is extended against resistance or if it is adducted across the palm, thereby increasing tension within the sheath, pain is reproduced. Tenderness and swelling over the tendon at the radial styloid are detected. Pinch and grasp are often weak.

Treatment. Conservative treatment consists of immobilization of the wrist and the thumb in a small cast for several weeks. Failure to improve justifies surgical intervention. A transverse incision is made over the radial styloid. The superficial fascia is split longitudinally with due care to avoid the superficial branches of the radial nerve in the deep aspect of the fascia. The sheath is cut or may be excised completely. If a third tendon is present, it is the aberrant tendon of the thumb and should be removed.

When the flexor pollicis longus tendon is severed, flexion of the distal phalanx is lost, and the proximal tendon retracts as high as the wrist. It is exposed in the thumb by a mid-lateral incision. In the thenar area, an incision parallels the thenar crease exposing the tendon deeply between the two heads of insertion of the flexor pollicis brevis. At the proximal part of the thenar prominence, the transverse carpal ligament must be severed and the motor thenar nerve protected against injury.

Trigger Thumb

In the act of grasp, the flexor pollicis longus tendon and sheath are compressed against a prominence of the head of the 1st metacarpal and the sesamoids in the heads of insertion of the brevis. Repeated trauma thickens the sheath, which in turn constricts the tendon. The traumatized tendon forms a bulbous enlargement which meets with momentary obstruction when passing through the stenosed sheath. Movement of the tendon enlargement through the narrowed canal produces a characteristic thump. Eventually, enlargement of tendon and sheath progresses to the point where movement is blocked and the thumb is locked, usually in flexion, so that the bulbous tendon remains proximal to the point of constriction. When the metacarpal head is unusually enlarged by spur formation, pain in gripping is an associated symptom.

Treatment consists of immobilization of hand and thumb in a cast for several weeks. Rest often will allow swelling to subside, and free motion will be restored. Stubborn cases are corrected by surgery. The constricted sheath is severed medially and laterally. Removal of sesamoids or any bony overgrowth eliminates the prominence and prevents recurrence. Occasionally, the condition is seen in infants in whom the tendon displays a diffuse enlargement where it passes through a normally thickened tendon sheath. Treatment is similar to that in adults, except that removal of sesamoids is unnecessary.

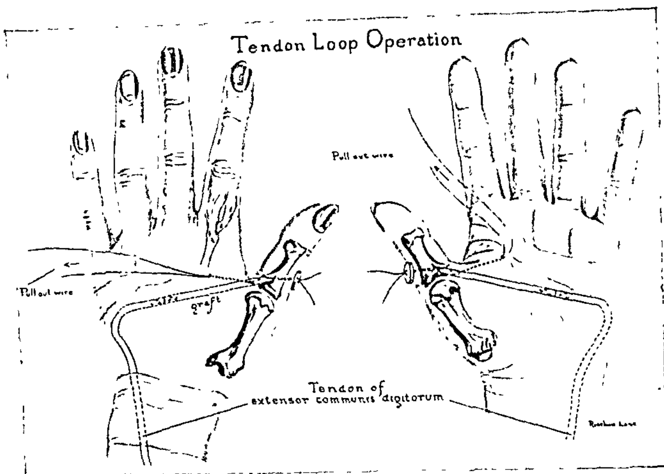


Fig. 334. In the tendon loop operation to restore adduction to the thumb, the extensor communis tendon from the index finger is prolonged with a tendon graft, and then passed subcutaneously around the ulnar border of the hand and across the palm under the flexor tendons to act as an adductor of the thumb. Attachment to the thumb is by fine stainless-steel wire through a drill hole to a button outside the skin. A flake of bone is chipped up at the insertion. A pull-out wire is placed so that the suture may be removed in 3 weeks. The stump of the extensor communis of the index finger is attached to the extensor indicis proprius to prevent rotation deformity. (Bunnell, S.: Surgery of the Hand, ed. 3, Philadelphia, Lippincott)

Operation to Restore Thumb Adduction and to Restore Arches. An extensor communis tendon is extended by a tendon graft, routed about the ulnar side of the hand, carried across the palm beneath the deep flexor tendons and attached to the base of the proximal phalanx of the thumb

An alternative procedure, which in addition to providing thumb adduction also restores the arches, is the Tendon T Operation. A tendon graft spans the palm between the base of the proximal phalanx of the thumb and the neck of the 5th metacarpal. A tendon transfer attached to the middle of this pulls it proximally into a V, thereby approximating the thumb and the little fingers and reforming the metacarpal arch.

Opposition of the Thumb

This is the most important motion in the hand. By it the thumb is brought forward across the palm and is rotated more than 90° so that the pulp faces that of the fingers, and the nail parallels the palm. Functions of grasp and pinch are effected. The opponens muscle which ensheaths the first metacarpal and is innervated by the thenar branch of the median nerve produces this movement. The flexor brevis, principally through its outer head inserting on the radial side of the proximal phalanx, aids the motion by flexing at the metacarpophalangeal joint and pronating the thumb The 1st metacarpal sits on a saddle joint and glides forward during the motion. Stabilization of the thumb at its 3 joints is a

The *abductor digiti quinti* acts like an intrinsic, attaching to the lateral tubercle of the proximal phalanx, contributing lateral fibers to a dorsal hood and forming a lateral band. Therefore, it flexes the proximal phalanx and abducts the finger. However, it has no extensor power on the distal phalanges. The opponens digiti quinti opposes the little finger. The thenar and the hypothenar muscles form the carpal arches.

Abduction and adduction of fingers can occur only with proximal phalanges extended. In this position, the collateral ligaments are loose. As the phalanx flexes downward over a wider volar articular surface of the metacarpal head, the collateral ligaments are diverted by wider bone and therefore tighten, preventing lateral motion. The metacarpals are anatomically in a divergent arrangement so that a slightly spread position of the fingers, when fully extended, should not be mistaken for intrinsic action.

Clinical Appearance of Intrinsic Loss

The middle and the proximal phalanges remain fully extended, and the distal phalanges are in partial flexion The fingertips can be flexed to the bases of the fingers by the long flexors but not to the palm. If the proximal phalanges are flexed passively, the distal phalanges cannot be extended actively. Lateral motion of fingers is lost The thumb cannot be adducted or opposed across the palm. Carpal and metacarpal arches are flattened A typical clawhand results, the fingers are clawed, the hand is flat, the thumb is at the side.

When the middle extensor slip to the base of the middle phalanx is ruptured, a "boutonnière" deformity results. The middle joint is flexed, and the lateral bands spread and displace volarward, the synovium herniating dorsally The lateral bands then act as flexors at this joint.

Avulsion of the distal insertion of the extensor aponeurosis results in a dropped fingertip, with loss of extension of this phalanx.

When the anterior capsule of the middle joint is ruptured, this joint becomes hyperextended, and the distal phalanx is drawn into flexion by the flexor profundus.

Treatment of Intrinsic Paralysis

Nerve suture is attempted early, before the muscles have undergone fibrous change. After this time, tendon transfers are required. This is preceded by mobilization and correction of deformities by traction, osteotomies, capsulotomies, etc. The dorsal aponeurosis is very thin and delicate, and repair is generally followed by adherence to the underlying bone. A simple stainless steel removable wire is used.

Operation. The flexor sublimis tendon is severed at its insertion and withdrawn into the palm. It is split longitudinally into 2 strands. Each strand is passed down a lumbrical canal and placed over the lateral band, which is scraped and to which the strand will adhere. A removable running wire suture fixes the tendon to the lateral band and each end passes through the skin where it is held by lead shot, and removed later.

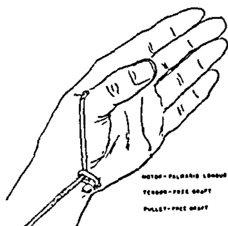
The strands from each sublimis should go to corresponding sides of fingers, either all to the radial side or all to ulnar side so that pull of this muscle pulls both fingers in the same lateral direction. Each finger should have a tendon for both sides to furnish both abduction and adduction. Available are the sublimis tendons, the 2 extensor proprius tendons and the palmaris longus. Enough tendons for thumb opposition and adduction should be reserved.

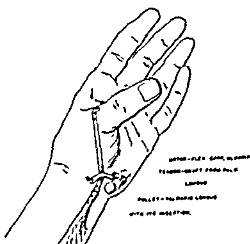
Severe Adduction Deformity of Index or Little Fingers

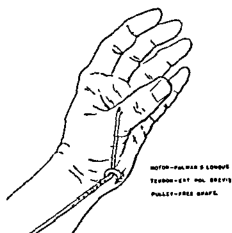
This is due to localized loss of the first dorsal interosseous or the abductor digiti quinti, respectively. The extensor indicis propius or the extensor digiti quinti proprius is inserted into the lateral lumbrical canal.

Thumb Adduction

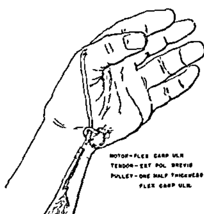
When the thumb adductor is paralyzed, the thumb cannot make a perfect "O" with the index finger and pinch between them is weak; the metacarpophalangeal joint falls into hyperextension because of loss of flexor power at this joint. The thumb cannot be scraped across the palm. Muscle atrophy in the first interosseous space is obvious The carpal and the metacarpal arches enable the fingers to converge on flexion and cup the hand, which is essential for grasping small round objects. These arches are absent when thenar, hypothenar and interosseous muscles are paralyzed.











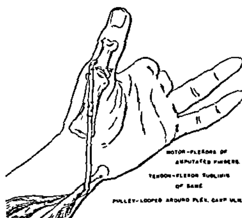


Fig. 336. Operation advised to restore ability to oppose the thumb. The 2 essential principles are: (1) the tendon should pull in the right direction; namely, subcutaneously diagonally across the thenar eminence toward the pisiform bone to angulate the thumb forward and toward the ulna; (2) the insertion of this tendon should be into the dorso-ulnar aspect of the base of the proximal phalanx, so as to pronate the thumb. Then one may use whatever seems best in the individual case for motor power, tendon and pulley. These drawings (*left, top to bottom; right, top to bottom*) illustrate what muscle and tendon material may be used, and ways of constructing the pulley. (Bunnell, S.: Opposition of the thumb, J. Bone & Joint Surg. 20:274)

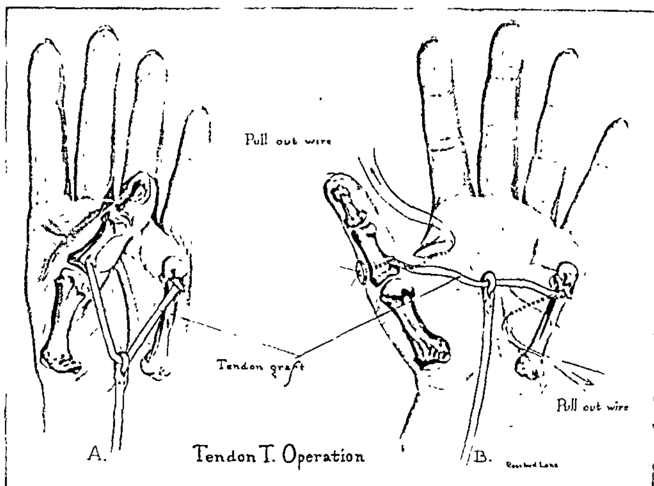


Fig. 335. In the tendon T operation to restore adduction to the thumb and curvature to the carpal and the metacarpal arches, a tendon graft spans the distance between the little finger metacarpal and the adductor insertion in the proximal phalanx of the thumb. A long flexor tendon of the forearm (a sublimis or the palmaris longus prolonged by a strip of its palmar fascia) is looped over its center to form the T (B). This, when in action, changes to a Y (A), adducting the thumb and curving the arches. (Bunnell, S.: Surgery of the Hand, ed. 3, Philadelphia, Lippincott)

necessary prerequisite for opposition This is accomplished at the carpometacarpal joint by the abductor longus; at the metacarpophalangeal joint by the extensor brevis, and at the interphalangeal joint by the long extensor. Loss of opposition is due to (1) paralysis of muscles or radial half of the thenar eminence, (2) cicatricial contracture between first 2 metacarpals, (3) fixation of extensor longus by adhesions, (4) loss of stabilizing muscles, and (5) ankylosis of intercarpal and carpometacarpal joints.

Surgical Restoration of Opposition

1. Nerve suture—the thenar branch
2. Removal of cicatrix—this frees the metacarpals.
3. Freeing of extensor pollicis longus
4. Bone operation

A. Rotary angulatory osteotomy of first metacarpal done at the base.

B. Carpal osteotomy done when carpals are ankylosed. A wedge is removed from the radial aspect.

C. Bone graft bridging first 2 metacarpals (Thompson, Allen, Foerster). This is used in cases with insufficient muscles and for excessive contracture in the first metacarpal space.

5. Tendon transference. This provides active opposition. The muscle selected should be strong, the pull should be toward the pisiform bone, and the insertion should be on the dorsal ulnar aspect of the base of the proximal phalanx. A preliminary rotary angulatory

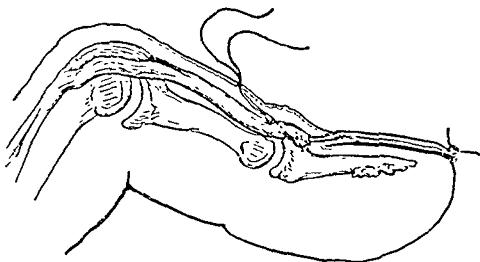


Fig. 338. Method of using stainless-steel wire for late repair of insertion of the extensor tendon, fastening the suture wire to the fingernail and later removing it by the pull-out wire. (Bunnell, S.: Primary repair of severed tendons: use of stainless-steel wire, Am. J. Surg. 47: 505)

be instituted within the first few hours before bacterial contamination spreads. General or block anesthesia is desirable. A tourniquet is applied. The wound is covered with sterile gauze, and the surrounding skin is scrubbed with soap and water and painted with antiseptic solution. Débridement is performed by lifting out all foreign matter and loose particles, irrigating freely with normal saline, and excising the entire surface of the wound down to normal-appearing tissue. Essential structures, including tendons, nerves, blood vessels, and articular surfaces are spared. Strong chemical antiseptics are destructive and should be avoided. Foreign bodies, including catgut, cause scar tissue reaction; therefore, ligation of bleeders only by very fine plain catgut is permitted. Oozing is controlled by compression. If oozing continues, a temporary tiny rubber drain is inserted. Then, reconstruction of essential structures is done, using very fine stainless steel wire which is reactionless. Fractures can be fastened with thin wire and tendons united. After this the skin is closed. If a defect exists, the adjacent area is incised, and a flap of skin is slid over, the donor defect being covered with a split-skin graft. *Covering of the deep avascular structures is essential;* if necessary, it should be done by immediate split-skin grafting. Compression dressings to reduce postoperative edema should be applied with care so as not to cause ischemia. A splint of plaster or a universal hand splint is applied in a position of function. Combined tetanus-gas bacillus antitoxin or a booster dose of tetanus toxoid is administered. The patient is placed at bed rest, the extremity is elevated, and antibiotics are started.

If the injured hand is seen quite late, or if the type of injury predisposes strongly to infection, the wound after the initial care should be left open and closed secondarily several days later after granulation tissue has formed. A tendon never should be sutured after 24 hours, because infection within the tendon sheath will slough the tendon and spread rapidly.

TENDON RUPTURE

A normal tendon when subjected to extreme tension will rupture (1) at its insertion, sometimes with avulsion of a flake of bone, (2) at the musculotendinous junction, and (3) rarely at the muscle origin. Rupture within the tendon itself occurs when tendon pathology exists, such as (1) swelling and softening which occur in the few weeks following a direct crushing injury, (2) chronic infection as tuberculosis and syphilis, (3) acute suppurative infection, (4) degeneration of advancing age, (5) tumor, and (6) attrition by friction over a bony prominence or irregularity.

Ruptured Extensor at Distal Phalanx (Baseball Finger; Mallet Finger)

Sudden forceful flexion of the distal phalanx, such as by a blow on the fingertip while the finger is fully extended, ruptures the joint capsule at the tendon insertion. A small piece of bone may be avulsed. Active extension of the finger is lost. Conservative treatment started early is effective. A plaster splint is

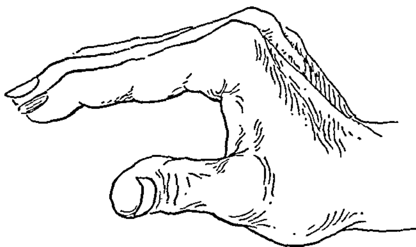


FIG. 337. Intrinsic contracture of the hand. The deformity corresponds to overaction of the intrinsics, namely flexion at the M-P joints and extension at the I-P joints. In addition, the thumb is adducted into the palm or against the index finger.

osteotomy may be indicated. Any available flexor muscle is selected. Most commonly used are the flexor carpi ulnaris, the palmar longus and the flexor digitorum sublimis of the ring finger. A pulley is constructed by a free tendon graft which is looped about the short muscle and the tendon attachment to the pisiform bone and is sutured to itself; or one half of the width of the flexor carpi ulnaris tendon may be used, suturing the free end to the pisiform bone. The extensor pollicis brevis is detached high, drawn out at its insertion, then passed subcutaneously across the palm, then through the pulley and is sutured to the distal end of the flexor carpi ulnaris. The normal function of ulnar volar flexion is not lost by this procedure. Instead, the palmaris longus may be used.

Intrinsic Contracture of the Hand; Surgical Treatment[2]

Clinical Picture. Metacarpal joints are strongly flexed, metacarpal arch is exaggerated, and interphalangeal joints are in full extension. This position corresponds to the picture of overaction of the intrinsics. It is opposite to the deformity of underaction, or paralysis, of the intrinsics, namely, extension at the metacarpophalangeal and flexion at the interphalangeal joints (clawhand of median and ulnar nerve paralysis and Volkmann's contracture). The thumb is also involved in the intrinsic contracture, being similarly flexed and extended, but also is adducted into the palm or against the index finger. This is the "intrinsic plus" hand of Bunnell in contrast with the "intrinsic minus" of the paralytic.

Etiology

1. Ischemia (constricting cast; vascular injury)
2. Spasm persisting (rheumatoid arthritis; Hansen's disease)
3. Fibrosis (thermal or compression injury)

Functional loss involves inability to flex the fingers unless the metacarpophalangeal joints are hyperflexed. Pinch and grasp require that the metacarpophalangeal joints be flexed about 45° and the interphalangeal joints partially flexed and the thumb opposed. Therefore, pinch and grasp are impossible. (Bunnell states that grasp is possible, but the hand cannot be opened fully before grasping.)

Clinical Test for Intrinsic Contracture. If the metacarpophalangeal joints are extended, the interphalangeal joints cannot be flexed.

Surgical Treatment. The extensor component in the interphalangeal joints must be released while maintaining the flexor component to the metacarpophalangeal joints. At the level of the metacarpophalangeal joint, the oblique fibers which form the lateral bands are excised through a mid-line incision over the dorsal aspect of the proximal phalanx. Postoperatively, the joint is splinted in full extension, and the interphalangeal joints are exercised actively. Bunnell strips the interossei from the metacarpals and displaces them distally. In addition, the collateral ligaments and the volar capsular ligaments are cut.

TREATMENT OF HAND INJURIES

The early care of an open wound of the hand is essential in preventing infection and subsequent severe crippling. Treatment should

[2] Harris, C., Jr., and Riordan, D. C.: Intrinsic contracture of the hand and its surgical treatment, J. Bone & Joint Surg 36A:10, 1954.

Trigger Finger

A fusiform swelling of the sublimis tendon at its bifurcation passes through a thickened and constricted sheath over the metacarpal head with difficulty. On attempting flexion or extension, the sudden escape of the nodule from the narrowed canal is accompanied by an audible snap, and the movement is completed. Eventually, the disproportion between tendon and sheath is too great to permit passage, and the finger becomes locked, usually in flexion. Trauma of compressing the tendon against the bony prominence in grasping is presumably the cause. In the thumb, the sesamoids underlying the flexor longus form a prominence over which the tendon is irritated. Spurs from an osteoarthritic metacarpal head frequently predisposes to this condition.

Acute cases gradually subside with rest. Prolonged cases require cutting the annular ligament laterally—never directly over the gliding surface. Underlying prominent sesamoid or spurs should be removed.

CHRONIC INFECTION OF THE HAND

Tuberculosis of the Hand

Tuberculosis in the bony structures is more common in children, is hematogenous, and secondarily involves the joint. The infection occurs in the medullary cavity of the phal-

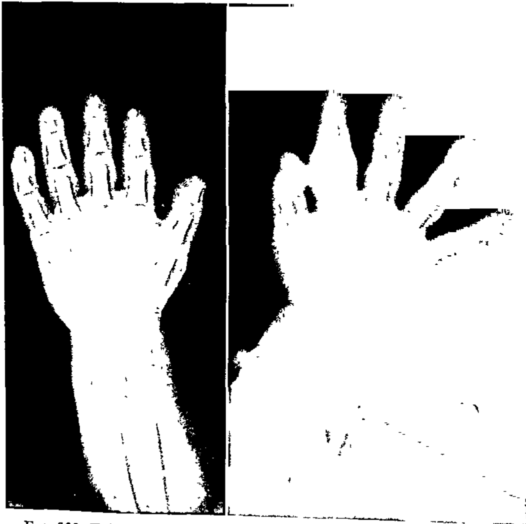


Fig 339. Tuberculosis of bone: (*left*) metacarpal; (*right*) phalanx. Periosteal reactive bone formation represents the healing stage. Before this takes place, the phalanx or metacarpal undergoes extreme decalcification and destruction by the tuberculous process.

applied for a period of 5 weeks with the middle joint flexed and the distal one hyperextended. After the 5th week, suture is indicated. Delicate handling of the thin aponeurosis is mandatory; otherwise, adhesions will form, and movement is lost. Number 35 stainless steel wire with pull-out wire attached is spliced into the tendon and drawn through the fingernail. If the tendon is united in a lengthened condition, it is shortened, underlying adhesions are freed, and the tendon is sutured with wire.

Rupture of Extensor at Middle Phalanx

A forced flexion injury avulses the middle slip from the base of the middle phalanx Occasionally, a direct injury to the tendon weakens it, and the tendon separates several days later. The middle joint goes into flexion, and the 2 lateral bands displace volarward. The joint capsule herniates through the opening. Early treatment consists of immobilization of the joint in hyperextension. When seen after a week, surgical repair is indicated.

Rupture of Extensor Pollicis Longus

This tendon runs through a groove on the dorsum of the radius, where under cover of the annular ligament it is subject to friction. At the distal edge of the dorsal carpal ligament, the tendon angulates sharply when the hand is dorsiflexed. Therefore, certain occupations requiring excessive use of the thumb and repeated dorsiflexion of the wrist encourage attrition of the tendon. A healed Colles' fracture produces an irregularity of the styloid and imposes added friction on the tendon. Clinically, there is inability to extend the distal phalanx of the thumb and to pinch the thumb against the side of the palm. Surgical repair often necessitates a free tendon graft to replace the damaged portion of tendon. An alternative is to transfer the extensor indicis proprius.

Rupture of Flexor Tendon

This is rare but may occur at the insertion of sublimis and profundus, over the lower end of the radius, particularly after a Colles' fracture, and at the musculotendinous juncture.

DISLOCATION OF TENDON

When the lower end of the ulna is torn loose from the radius, as in a Colles' fracture, and a typical Madelung's deformity results, the dorsal carpal ligament may be torn, and the extensor carpi ulnaris tendon dislocates over the ulnar styloid during pronation.

Not infrequently, the common extensor tendon, particularly that to the index or the middle fingers, may dislocate ulnarward from its position over the metacarpophalangeal joint when the joint is flexed. Attempted extension of the fingers thereafter is limited; instead, the finger moves ulnarward. When the finger is extended passively, the tendon slips back to its normal position. The cause presumably is rupture of the radial aspect of the joint capsule. Repair of the capsule is indicated.

TRAUMATIC TENOSYNOVITIS

Direct trauma to, or excessive use of, a tendon and its synovial sheath causes synovial inflammation. The synovial surface becomes dry and covered with fibrin. Movement of a tendon within an enclosure lacking lubrication produces clinically detectable crepitation. The tendon is likely to be irritated where it passes over a bony prominence such as the radial styloid in DeQuervain's disease, or the sesamoids at the base of the thumb in a trigger thumb. Inflammation is greatest at a musculotendinous junction when the muscle is overstrained.

Clinically, pain is experienced over the course of muscle and tendon and is accentuated by active or passive motion of the tendon. The tendon sheath is tender, and crepitation is detected with movement.

Treatment in mild cases consists of avoiding the offending movement, resting the part and applying hot moist packs. Gentle exercises are advisable to prevent adhesion formation. Severe cases may be splinted temporarily until the acute symptoms have subsided.

One must be exceedingly careful to rule out a low-grade infectious process or gout before classifying a tendinous involvement as a traumatic tenosynovitis. The situation is clarified when inflammatory signs in the skin become apparent.

and chemotherapy. Function is better after radical excision. If the tendon is ragged, it should be removed and tendon grafting done. If excision and after-care fail to produce the desired result, particularly if osseous involvement has occurred, excision of the infected bone and arthrodesis are indicated.

Syphilis of the Hand

In the tertiary stage the skin, the tendon sheaths, the bones and the joints may be involved. In bones, proliferation of periosteal bone and greatly increased density are typical. Destructive areas occur throughout the diaphysis. The finger, as in tuberculosis, is swollen, boggy and spindle-shaped. Gummatous osteomyelitis may break down and cause discharging sinuses and sequestrum formation. Joint ankylosis occurs. Early wrist joint involvement displays a chronic painful synovitis with effusion. Later, a gummatous synovitis with destruction of cartilage and bone results. The painless destruction of a Charcot joint is another entity. This may occur in the wrist and the metacarpophalangeal joint.

DUPUYTREN'S CONTRACTURE[3-6]

Dupuytren's contracture consists of hypertrophy and contracture of the palmar aponeurosis and consequent flexion deformity of the distal palm and fingers.

Etiology

The exciting cause is unknown. Antecedent trauma and manual labor have no relationship. A familial tendency is often noted. The condition starts in adulthood, usually after the age of 30. Frequently, it is associated with fibrous contractures elsewhere in the body, particularly in the plantar fascia.

Pathology

The area of the distal palm proximal to the ring and the index fingers is involved most commonly. Early, the palmar aponeurosis at this site thickens. Microscopically, immature fibroblasts and round cells are found. The overlying subcutaneous fat disappears, and the skin becomes intimately attached to the fascia. An abundance of fibrous tissue is formed which grossly appears as nodules. The process extends distally into the digital prolongations of the aponeurosis. The tendons, the vessels and the nerves themselves are not involved. Secondarily, the proximal interphalangeal joint undergoes a flexion contracture.

Clinical Picture

The condition is frequently bilateral and symmetric. The course is chronic, taking anywhere from 1 to 20 years to reach the maximum deformity. It usually starts with a nodule in the palm at the distal palmar crease proximal to the ring finger. A fibrous cord develops proximally and distally, and the finger is pulled into flexion at the metacarpophalangeal and the proximal interphalangeal joints. The aponeurotic prolongation reaches only as far as the middle phalanx and explains the freedom from involvement of the distal phalanx. The ring and the little fingers are involved most commonly; less commonly, the long finger; and rarely, the index finger and the thumb. The palmar mass is composed of distinct bands or a dense plaque fused to the overlying skin, which is thick and indurated. Symptoms are not marked. The main complaint is deformity and the interference of the flexed fingers on use of the hand.

Treatment

Correction of the deformity is surgical. The palmar aponeurosis and its digital prolongations are removed.

Surgical Technic. A tourniquet is applied. A transverse incision is made across the palm in the distal palmar crease. This may be extended proximally at the ulnar border of the hand. At the proximal part of the palm, the aponeurosis is severed from the transverse carpal ligament and drawn distally. At the level of the proximal palmar crease and just distal to the superficial palmar arch, the aponeurosis sends vertically disposed sheets of fascia deeply on both sides of each metacarpal

[3] Bunnell, S.: Surgery of the Hand, ed. 3, Philadelphia, Lippincott, 1956

[4] Mason, M. L.: Arch Surg 65 457, 1952

[5] Richards, H. J.: Surgical treatment of Dupuytren's contracture, J. Bone & Joint Surg. 36B:90, 1954.

[6] Tubiana, R.: Prognosis and treatment of Dupuytren's contracture, J Bone & Joint Surg 37A:1155, 1955.

anges, causing patchy destruction. The periosteum becomes distended with granulation tissue, forms a bony involucrum, and the diaphysis sequestrates. In adults, the infection is mainly subperiosteal, shutting off the blood supply to the shaft which immediately necroses and is extruded before a bony involucrum can form. As a result, the finger is shortened The course is chronic. A cold abscess forms and may rupture; persistent draining and secondarily infected sinuses result. The infected finger joints become ankylosed The spindle-shaped distended bone is characteristic. Treatment consists of curetting the bone, closure without drainage, prolonged immobilization, and antibiotics and chemotherapeutic agents specific for tuberculosis. The prognosis is good only in children Obstinate drainage and crippling of the finger justifies amputation

Tuberculosis of the wrist joint is more common in children, in whom it is secondary to carpal infection. In adults, it results from spread of an adjacent tenosynovitis. The wrist exhibits a doughy, fusiform swelling but little or no redness. The muscles of the forearm are atrophic They may show spasm. Roentgenograms show osteoporosis and destruction of the carpal bones. Eventually, a cold abscess and sinuses form. Treatment consists, in early cases, of prolonged immobilization In advanced cases, the carpal area is resected, closed without drainage and immobilized; then antibiotics and chemotherapy are started.

Tuberculosis tenosynovitis is an insidiously developing progressive swelling and inflammation of the tendon sheath with pain and limitation of motion. People who work with cattle seem to be predisposed, the disease usually being of the bovine type. The tendons of the wrist are not commonly involved It is rarely associated with pulmonary tuberculosis. It usually starts as a simple inflammation of the tendon sheath with serous exudate and fibrinous deposits covering the inner lining The sheath thickens and becomes granular and the exudate seropurulent. A sheath may undergo caseation, and the exudate penetrates to form sinuses. Or the sheath may heal and become fibrotic When the synovial lining is infected, the surrounding paratenon and fascial structures (palmar, thenar and quadrilateral spaces) are involved by contiguity. The infection spreads along the sheath to involve the area from muscle to tendinous insertion; it also *spreads to other tendons and all the fingers may be affected.* On the back of the hand all the extensors are involved, but there is no extension to the fingers.

The membrane is dull, red, villous, and covered with fibrin, which floats into the exudate as rice bodies. Granulation tissue spreads to the surrounding tissue, engulfing all structures Eventually, the granulation tissue invades the tendinous structure itself, which is destroyed or ruptures spontaneously. The muscle structures and nerves are fairly resistant to invasion. The infection may penetrate ligaments and eventually invade the wrist joint and the carpal bones. The melon-seed-shaped rice bodies are composed of laminated layers of fibrin.

The course is slow and insidious, and symptoms over several years are mild Complaints include swelling, pain, tenderness, weakness of grasp, and limitation of motion. Swelling commonly occurs above the wrist as an elongated doughy mass extending to one side of the palm and narrowed at the level of the transverse carpal ligaments The swelling then extends down to a finger, most commonly the middle finger. The swelling is soft and doughy; in the early stage it is movable. The overlying skin is normal or at the most presents a slight edema or redness. Induration is an advanced sign. The early fibrinous stage displays creaking and crepitations on motion. As exudate accumulates and rice bodies form, fluctuation and these loose bodies are perceptible. Pain and tenderness are mild. Stiffness and weakness complaints are usual. Limitation of motion early is nil but gradually increases with progressive adherence of the tendons. Fever and general symptoms are absent. Spontaneous rupture of a tendon may occur. Sinuses are late complications The finger and the wrist joints and bony structures may be invaded.

Extreme conservatism is indicated in the acute spreading stage. Immobilization until the process is reduced in severity is followed by resection of all diseased tissue, primary closure, continued immobilization, antibiotics

latter time. Free split-thickness grafts are inadequate.

Conservative Treatment. Massive doses of vitamin E have been used with questionable results.

CALCIFIC DEPOSITS IN THE HAND AND THE WRIST[7-10]

Amorphous calcium salts deposited in the soft tissues, particularly about the wrist, may be the cause of severe pain of sudden onset. The condition is similar to that seen more commonly in the shoulder. Clinically, the patient holds the wrist and the hand protectively rigid; any motion causes severe pain. The wrist is swollen, and the tissues are reddened, indurated and exquisitely sensitive. The roentgenogram shows an opacity in the soft tissues, particularly about peritendinous structures. The site of insertion of the flexor ulnaris is frequently involved. The process is usually self-limited, the deposits disappearing in a few weeks' time. To alleviate pain, procaine injections and immobilization in a cast are effective.

Sutro and Cohen suggest an underlying vitamin deficiency (particularly of vitamin E) as the cause. Degeneration, necrosis and calcification of voluntary muscle fibers develop in animals kept on a diet deficient in vitamin E.

CONGENITAL ARTERIOVENOUS FISTULA[11]

Congenital arteriovenous fistula consists of multitudinous abnormal communications between an artery and a vein proximal to the capillary bed. Theoretically, it arises from failure of the embryologic anlage to differentiate into separate artery and vein. Normally, both artery and vein differentiate from a common capillary plexus, and the communications between them are ordinarily obliterated. In this condition, they persist, and the blood flow is short-circuited.

Clinical Picture

1. *The hand and the forearm are often involved.* An entire or limited portion of either the upper or the lower extremity may be affected.
2. *Varicose veins* and concomitant ulceration at the distal parts of the extremity
3. *Gangrene* of the tip of the finger is due to ischemia.
4. *Birthmarks* in more than half of cases. Consist of port-wine hemangiomas or diffuse hemangiomas.
5. *Hyperhidrosis and hypertrichosis* in the affected area
6. *Increased growth of limb.* Increased length
7. *Higher skin temperature*
8. *Thrills and bruit uncommon* in contrast with the acquired type of arteriovenous fistula in which the communication is large.
9. *Branham's bradycardia sign.* Reduction in the heart rate occurs when the fistulous area is obliterated by compression. This is more common in the acquired type.
10. *Increased oxygenation of venous blood.* Blood drawn from a vein proximal to the communication channel may be as much as 45 per cent greater than that from the opposite extremity.

Arteriography

This demonstrates the site of communications and the completeness of surgical eradication. A sphygmomanometer is inflated well above systolic pressure, and 30 cu. cm. of Diodrast is injected into the brachial artery. An x-ray exposure is made. Additional films are taken at systolic pressure, diastolic pressure, and at zero. The following are characteristic findings:

1. Dilatation of arteries leading to the fistulae
2. Absence of normal filling of arteries distal to the fistulae
3. Pooling of the arteriographic medium in the region of the fistulae. The pattern made

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[7] Lapidus, P. W. Infiltration therapy of acute tendinitis with calcification, Surg, Gynec. & Obst 76 715, 1943.

[8] Milch, H, and Green, H. H.: Calcification about the flexor carpi ulnaris tendon, Arch. Surg. 36.660, 1938

[9] Serdenstein, H· Acute pain in the wrist and hand associated with calcific deposits, J Bone & Joint Surg 32A 413, 1950

[10] Sutro, C. J., and Cohen, L. J.: Basis and treatment of calcification of tendinocapsular tissues, especially the supraspinatus tendon, Arch. Surg. 42:1065, 1941.

[11] Curtis, R M: Congenital arteriovenous fistulae of the hand, J. Bone & Joint Surg 35A.917, 1953.

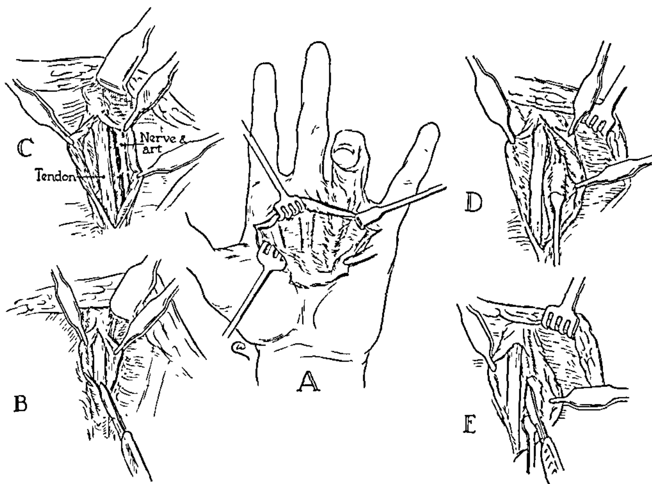


Fig. 340. Dupuytren's contracture, surgical correction. Note that the neurovascular bundle and the lumbrical muscle lie in a vertical septum which extends deeply from the palmar aponeurosis to the deep palmar fascia and the side of the metacarpal. This vertical fascia must be dissected out and removed. (Mason, M. L.: A.M.A. Arch. Surg. 65:457)

to attach to the deep palmar fascia and the sides of the bone. These septa enclose the tendons. Within their substance, these septa split to form secondary channels which enclose the neurovascular bundles and the lumbrical muscles. As the aponeurosis is elevated, the vertical septa are severed deeply, the digital vessels and nerves are dissected out, the lumbrical muscles are exposed in their canals, and thus the fascia are freed further distally. Next, through a mid-lateral longitudinal incision in the finger, the dissection and removal of the fascia are completed. This last step permits extension of the fingers. Full extension may not be gained because of secondarily contracted structures. The tourniquet is released, and complete hemostasis secured. The skin is closed, and drains are inserted to be removed in 48 hours. Hydrocortone solution injected into the wounds aids in reducing postoperative reaction and fibrosis. A compression dressing is applied, and the hand and the fingers are bandaged in flexion. No attempt should be made to gain immediate extension, as this puts too much tension on the skin, causing it to necrose and slough. The distal phalanges of the fingers are left free for immediate gentle exercises. The drains are removed in 48 hours. Immobilization is discontinued at 2 weeks, and exercises are started. A splint may be applied to obtain gradual extension of the affected fingers. The fingers and the palm straighten over a period varying from a few months to 2 years, and the skin gradually thickens and acquires a subcutaneous fat pad.

A skin graft is rarely required. When necessary, a tubular pedicle graft is attached to the ulnar side of the hand 3 weeks prior to surgery, and the transfer is completed at the

are seen throughout. The giant cells are frequent, numbering as many as 30 per high power field. These cells possess a faintly eosinophilic cytoplasm which is foamy, due to lipoid droplets and hemosiderin deposits. Nuclei vary from 3 to 100. Foam cells are not constant and are displayed best near the periphery. Their nuclei are small and stain more darkly than the polygonal type cells. They give the tumor the *typical bright yellow color*. The hemosiderin pigment occurs mainly in the phagocytes, occasionally in the giant and foam cells. Small cystlike endothelial-lined spaces are seen, suggesting synovium. The small vessels show evidence of endarteritis and partial obliteration of their lumens This suggests an inflammatory etiology. Tendon sheath tumors are more fibrosed and less vascular than joint or bursal xanthomas. They are richer in lipoid but less pigmented with hemosiderin.

Clinical Picture. Discrete single or multiple masses appear over the course of tendons of the hands and feet and the Achilles tendons. Bilateral involvement is usual. The tumors are discrete, their consistency varies from soft to firm, they are generally pea-sized although they may be large, bulbous, and disfiguring, and they are fixed to and move with the underlying tendons. The overlying skin is normal in appearance and is freely movable over the tumors. Tenderness is unusual. Growth is very slow and takes place over a number of years. The condition is often a part of hypercholesteremic xanthomatosis, a familial metabolic disturbance, and other lesions, such as yellow plaques about the periorbital soft tissues, may be evident. When the tendon tumors first make their appearance, they are painful, but later they are painless. Spontaneous tendon rupture is a rare complication.

Laboratory Findings. An elevated serum cholesterol is often associated.

Roentgenologic Findings. These reveal pressure erosion of the adjacent phalanx. The articular surfaces are never involved.

Treatment. Complete excision is advised. Reconstruction of the tendon by a tendon graft may be necessary. Recurrence is common.



FIG. 341. Arteriogram of congenital arteriovenous fistulae of the hand. 30 cc. of Diodrast injected into brachial artery. The contrast medium characteristically puddles in the thenar area in a snowflake pattern. A large superficial vein is visible along the radial artery. Volar digital arteries of the thumb do not fill. Terminal phalanx of thumb has "moth-eaten" appearance. (Curtis, R. M.: Congenital arteriovenous fistulae of the hand, J. Bone & Joint Surg. 35A:917)

by the contrast medium is suggestive of snowflakes in the region of the communications.

4. Appearance of the contrast medium in the veins distal to the point of injection while the proximal artery is occluded.

Treatment

The excision described for the acquired type (q.v.) is of no value in the congenital type. Gangrene often develops, and amputation becomes necessary. However, by planning *the excision in stages, adequate collateral circulation* may develop between procedures. The overlying skin should be replaced with a pedicle graft.

*A cavernous hemangioma should be re*moved as this may prevent the development of an arteriovenous fistula.

BENIGN GIANT CELL TUMOR OF TENDON SHEATH[12-15]
(Benign Synovioma; Xanthoma of Tendon Sheath)

This is a common tumor in the tendon sheath, particularly of the flexor tendons of the hand. It occurs predominantly in women in the 3rd and the 4th decades. It may also *occur on the extensor aspect of a finger where* it arises from the synovium of an interphalangeal joint. Rarely, the ankle and the toes are involved.

The tumor is pea-sized, seldom larger, firmly attached to the site of origin, bright yellow in color, encapsulated and characteristically lobulated. The cut section displays streaks of yellow and brown.

Microscopically, the tumor is very cellular. The matrix consists of collagenous connective tissue, which is largely hyalinized, thereby staining a brilliant pink with hematoxylin and eosin. Groups of polygonal cells with scanty *slightly basophilic cytoplasm and large round* or oval nuclei with light chromatin network

[12] Fletcher, A. G, Jr, and Horn, R C.: Giant cell tumors of tendon sheath origin, Ann Surg. 133: 374, 1951.

[13] Spencer, H, and Whimster, I W.: Development of giant-celled tendon-sheath tumors and related conditions (chronic villo-nodular synovitis and cutaneous histiocytoma), J. Path & Bact. 62:411, 1950.

[14] Wright, C. J. E.. Benign giant cell synovioma, Brit. J. Surg. 38 257, 1951.

[15] Jaffe, H L, Lichtenstein, L, and Sutro, C. J: Pigmented villonodular synovitis, bursitis, and tenosynovitis, Arch Path. 31:731, 1941.

The deep inguinal glands also drain the pelvic organs, often becoming painfully swollen in pelvic inflammatory disease.

Cutaneous Nerves. The *lateral*, the *intermediate* and the *medial cutaneous nerves of the thigh* (from the femoral) supply the lateral, the anterior and the medial aspects, respectively, of the thigh. They pierce the deep fascia along an oblique line that corresponds to the position of the sartorius. The lateral femoral cutaneous nerve often arises independently from the lumbar plexus and enters the thigh beneath the inguinal ligament close to the anterior superior iliac spine. It descends over the surface of the sartorius embedded in a ridge of thick fascia. It is involved in meralgia paresthetica, a condition characterized by sensory deficit along the lateral surface of the thigh.

The *ilio-inguinal nerve* (from lumbar plexus at L1) emerges from the superficial inguinal ring and supplies not only the scrotum or the labium majus but also the skin of the adjacent aspect of the thigh.

The *genitofemoral nerve* (from the lumbar plexus at L1, 2) sends a femoral branch to supply an area immediately below the inguinal ligament.

One should note that disease or trauma about the upper lumbar plexus produces symptoms referable to the front of the thigh; involvement of the lower plexus which forms the lumbosacral trunk will produce symptoms in the sciatic distribution.

Scarpa's Triangle (Femoral Triangle). This triangular area is bounded above by the inguinal ligament, laterally by the sartorius and medially by the medial border of the adductor longus. The flattened tendon of the adductor longus attaches to the front of the body of the pubis and runs laterally with the pectineus, forming the floor of the triangle. On passively abducting the thigh, the adductor longus is rendered taut and is palpable as a firm cord beneath the skin where it may easily be severed in performing an adductor tenotomy.

The femoral artery crosses the midinguinal point and enters the femoral triangle, then leaves beneath the sartorius within *Hunter's adductor canal.* It is within the triangle that the artery is most accessible in the thigh.

The head of the femur can be localized by determining the midinguinal point of crossing of the femoral artery, then measuring 1 inch distally and 1 inch laterally.

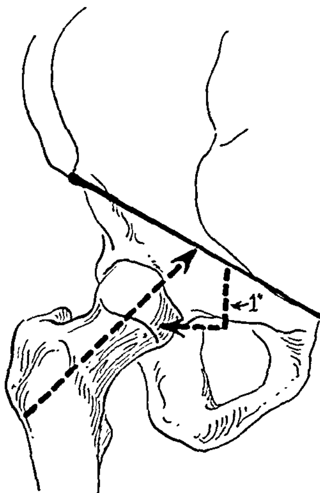


Fig. 342. Localization of the hip joint. *Femoral head:* from the point where the femoral artery crosses the inguinal ligament, measure 1 inch distally, then 1 inch laterally. *Femoral neck:* from a point at the base of the greater trochanter through a line intersecting the midpoint between the anterior superior iliac spine and the symphysis pubis.

The floor of the triangle is a trough whose medial sloping wall is formed by the pectineus and the adductor longus muscles, and the lateral wall by the iliopsoas and the vastus medialis.

Femoral Artery. This vessel runs behind the midinguinal point and enters the femoral *triangle in a trough between muscles supplied* by the obturator nerve medially, and by the femoral nerve laterally, except that the pectineus, lying among the adductors is supplied by the femoral nerve.

24

The Hip

SURGICAL ANATOMY

A. The Anterior Hip Region

Bony Landmarks. Palpable bony prominences include the *anterior superior iliac spine,* behind which at the widest part of the pelvis is the *tubercle of the iliac crest;* the *pubic symphysis* in the mid-line anteriorly; the *pubic tubercle* about 1 inch lateral to the symphysis; the *greater trochanter* of the femur; and the *pubic arch* felt at the uppermost limit of the medial aspect of the thigh. A line drawn horizontally from the pubic tubercle passes through the femoral head.

Fascia Lata. This is the thickened deep fascial covering of the thigh. It is attached above at the anterior superior iliac spine, the inguinal ligament, the body of the pubic bone, the pubic arch, the ischial tuberosity, the sacrotuberous ligament and, as the gluteal fascia, is attached to the sacrum and the iliac crest. It is extremely strong laterally because in between two layers of circularly disposed fibers there runs a broad band of coarse vertical fibers, the *iliotibial tract or band.* This tract is the conjoint aponeurosis of the tensor fasciae latae and the gluteus maximus and runs distally to insert upon the head of the fibula and the anterolateral aspect of the tibia. The band is often thickened and contracted in poliomyelitis Because the plane of the band lies lateral and anterior to the axis of hip motion, shortening of the band produces a flexion and abduction deformity of the hip

The fascia lata sends partitioning sheets inward to insert on the femur and separates the thigh extensors, the flexors and the adductors into groups.

Superficial Fascia. This is composed of a fatty layer and a deep membranous layer. The latter is separated from the deep fascia by loose areolar tissue but is firmly fused to the deep fascia along a horizontal line immediately below the inguinal ligament and to the front of the pubis and the pubic arch. Because of this attachment, urinary extravasations from the perineum to the abdomen cannot descend into the thigh.

The areolar tissue which covers the deep fascia is most pronounced over the anterolateral aspect of the lower thigh. This tissue is used for surrounding tendon junctures after repair to prevent adherence.

Long Saphenous Vein. This courses along the medial aspect of the thigh. It receives 3 tributaries which accompany the superficial inguinal branches of the femoral artery: (1) *superficial external pudendal,* (2) *superficial epigastric* and (3) *superficial circumflex iliac.* The last-named passes laterally below the inguinal ligament to the crest of the ilium where it is severed in anterior approaches to the hip. The long saphenous vein then passes through an opening in the fascia lata, the *fossa ovalis,* to reach the femoral vein. This latter aperture in the deep fascia is located 1½ inches below and lateral to the pubic tubercle where it is of surgical importance in treating femoral hernia or varicose veins. The fossa ovalis is covered by a thin layer of deep fascia, the *cribriform fascia,* behind which are located the femoral vessels.

Inguinal Lymph Glands. These consist of 2 groups. The *superficial group* consists of an *upper horizontal group* lying below the inguinal ligament and draining the inguinal, the lower abdominal and the perineal regions. The *lower group* lies about the long saphenous vein and receives superficial lymph vessels from the lower limb. The *deep group,* 1 to 3 in number, lies on the medial side of the femoral vein in the femoral canal and receives the deep lymph vessels of the lower limb, including those draining the popliteal glands.

from the medial and the lateral lips of the linea aspera. They have no action upon the hip.

The vastus lateralis at its proximal end is composed of a thick aponeurotic tissue which is attached to the base of the greater trochanter and is continuous above with the tendon of the gluteus medius. The descending branch of the lateral femoral circumflex artery runs along the anterior border of the muscle. In exposing the lateral aspect of the femoral shaft, incisions should be made along the posterior border and the muscle elevated forward to lessen the amount of bleeding.

Adductor Muscles. This group, which arise from the ascending ramus, the body and the descending ramus of the pubis and insert upon the linea aspera and the supracondylar ridge, function as adductors, flexors and lateral rotators of the thigh.

These muscles are disposed in 3 layers. The *anterior layer* is composed of the pectineus (femoral nerve) and the adductor longus (anterior branch of the obturator nerve). The *middle layer,* which is interposed between the anterior and the posterior divisions of the obturator nerve, consists of the adductor brevis (anterior branch of the obturator nerve). The *posterior layer* is the adductor magnus. This is a large fleshy muscle which arises mainly from the side of the pubic arch and is inserted along the back of the femur, being attached to the gluteal tuberosity, the linea aspera and the medial supracondylar ridge. This large muscle mass is innervated by the posterior branch of the obturator nerve and acts like other adductor muscles. However, the thick medial portion of the muscle arises from the ischial tuberosity and descends almost vertically to insert upon the adductor tubercle Like the hamstrings which also originate at the ischial tuberosity, this medial prominent muscle mass is innervated by the sciatic nerve and acts to extend the hip The dual nerve supply also explains the preservation of some adductor power after obturator neurectomy.

By passively abducting the hip joint, the adductor tendons, particularly that of the longus, are rendered taut and easily palpated and can be severed subcutaneously.

The anterior branch of the obturator nerve gives off an articular branch to the hip joint.

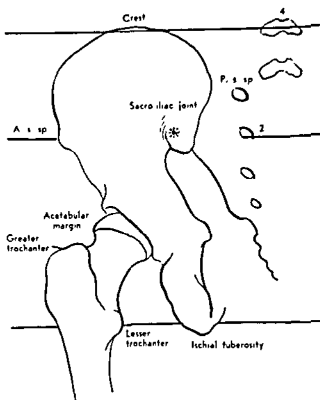


FIG. 343. Bony landmarks of the gluteal region. The uppermost portion of the iliac crest lies at the level of the 4th lumbar vertebra. The posterior superior iliac spine (P.s.sp.) lies at the level of the first sacral foramen, lower end of the dural sac, and middle of the sacro-iliac joint. A line drawn transversely across the ischial tuberosity crosses the lesser trochanter and indicates the interval between the quadratus femoris and the adductor magnus. (Redrawn from Grant: A Method of Anatomy, ed. 4, Baltimore, Williams & Wilkins)

This must be severed when denervating the joint. (See Obturator Nerve in the section on Neurology.)

Psoas Major and Iliacus. Both muscles originate intra-abdominally and enter the thigh behind the inguinal ligament. They are inserted by a common tendon into the lesser trochanter, which lies in a posteromedial location. The united iliopsoas crosses over the capsule of the hip joint, a bursa intervening between the structures. This bursa often communicates with the joint through an aperture in the capsule and is affected by the same disease processes that affect the joint.

Infections of the upper lumbar spine, particularly tuberculosis, often develop an exudate which descends downward beneath the

Profunda Femoral Artery. This arises from the lateral aspect of the femoral artery at or just below the level of the inguinal ligament. Rarely, it may arise as much as 4 inches below the ligament. It is a large vessel only slightly less in caliber than the femoral artery itself. It leaves the triangle by passing backward between the pectineus and the adductor longus muscles.

Femoral Circumflex Arteries. Two important large arteries spring from the profunda near its origin:

1. The *lateral femoral circumflex artery* runs laterally among the branches of the femoral nerve and passes between the sartorius and the rectus femoris muscles. It ends by dividing into 3 branches: a descending branch which courses along the anterior border of the vastus lateralis, a transverse branch, and an ascending branch which supplies the hip joint and anastomoses with the superior gluteal artery.

2. The *medial femoral circumflex artery* passes backward between the psoas and the pectineus muscles and ends at the upper border of the adductor magnus by dividing into 2 branches. The ascending branch enters the hip joint through the posterior capsule. The transverse branch ends in the hamstring muscles.

The branches of the lateral and the medial femoral circumflex arteries anastomose with each other, with the first perforating artery below, and with the gluteal arteries above to form the *crucial arterial anastomosis of the hip.* This system provides collateral circulation to the distal portions of the extremity in the event that a main vessel of the pelvis or the hip is occluded. Thus the ligation of the femoral artery, particularly in a young individual, is followed by minimal loss (e.g., toe) or none at all.

The course of the femoral artery lies along a line from the midinguinal point to the adductor tubercle.

Femoral Vein. This vessel lies medial to the femoral artery in the femoral sheath, but distally at the apex of the triangle it comes to lie behind the artery, as does the profunda artery and vein.

Femoral Nerve (L2, 3, 4). This nerve enters the thigh just lateral to the femoral artery and immediately breaks up into numerous branches. Muscular branches include those to the pectineus, the sartorius and the quadriceps. Cutaneous branches are the medial femoral cutaneous, the intermediate femoral cutaneous and the saphenous.

Two branches are closely applied to the lateral aspect of the femoral artery, accompanying the artery along the adductor canal: one supplies the vastus medialis; the other is the saphenous nerve. Only one branch passes to the medial side of the artery to supply the pectineus in spite of the latter's being an adductor. This explains why some adductor power remains after complete obturator neurectomy.

The femoral nerve lies in the groove between the iliacus and the psoas major where a large number of branches immediately cross the iliacus under cover of its fascia to reach the rectus femoris. In anterior hip-joint exposure, these branches must be secured and protected against injury.

Sartorius Muscle. This muscle arises by a fibrous origin with the inguinal ligament from the anterior superior iliac spine. In the adult, it extends distally for approximately 18 inches to insert into the anteromedial surface of the tibia below the level of the tubercle. It not only flexes the knee but flexes, abducts and externally rotates at the hip. Therefore, the muscle places the limb in a position assumed by a tailor while at work (*sartor* = tailor). It is supplied by the femoral nerve. The lateral, the intermediate and the medial femoral cutaneous nerves sometimes pierce it. The patellar branch of the femoral nerve always pierces it. The sartorius covers the roof of Hunter's canal and is superficial to the lower half of the femoral vessels.

Rectus Femoris. This muscle, supplied by the femoral nerve, arises by a straight head from the anterior inferior iliac spine and by an oblique head from the supra-acetabular rim. It not only reinforces quadriceps extension of the knee but also acts to flex the hip joint. Because it is innervated by the femoral nerve, it should be reflected medially in hip-joint exposures.

The 3 vasti muscles envelop the femur. The vastus intermedius arises from the anterior and the lateral aspects The vastus medialis and the vastus lateralis overlie the medial and the lateral aspects and arise by aponeuroses

mus, innervated by the inferior gluteal nerve, is a thick fibered rhomboid-shaped muscle, the superficial structure of the gluteal region. It arises from the posterior superior iliac spine, a small portion of the adjacent dorsum ilii, the sacrotuberous ligament, the lower sacrum and the coccyx. It passes laterally and distally toward the upper femur where a portion of its deep fibers insert into the gluteal tuberosity, the remainder ending in a band-like aponeurosis which joins a similar band-like aponeurosis of the tensor fascia latae, distal to the greater trochanter to form the *iliotibial tract*. Many muscle fibers through the iliotibial tract and the lateral intermuscular septum gain attachment to the linea aspera of the femur.

In the interval where the aponeurosis of the gluteus maximus plays across the surface of the greater trochanter is located a bursa. Another bursa is located below the lower edge of the muscle overlying the ischial tuberosity. A third bursa separates the maximus from the vastus lateralis.

The function of the gluteus maximus is to extend the hip joint. This motion is assisted by the hamstring tendons which attach to the ischial tuberosity. In the erect standing position, the gluteus maximus is relaxed.

Many arteries and large veins enter the undersurface and run transversely to the grain of the muscle. A muscle-splitting incision will cause extensive hemorrhage. The muscle should be freed laterally and elevated medially.

By severing the aponeurosis and reflecting the muscle medially, a layer of areolar tissue and fat lies beneath the muscle, and when cleared the remaining structures are exposed. The guide to these structures is the pyriformis, which emerges from the pelvis through the greater sciatic foramen and passes laterally to attach to the tip of the greater trochanter. All the main gluteal vessels and nerves also go out from the pelvis through the greater sciatic foramen.

The posterior cutaneous nerve of the thigh adheres ensheathed in fat to the deep surface of the maximus. First, it must be secured at the caudal edge of the muscle and freed as the muscle is raised.

Structures Entering at the Lower Border of the Pyriformis. These include the sciatic nerve, the posterior cutaneous nerve of the thigh, the inferior gluteal nerve and the inferior gluteal vessels. Also, the internal pudendal vessels and nerve, which emerge at the medial end of the lower border of the pyriformis and disappear immediately through the lesser sciatic foramen on their way to the perineum.

Structures Entering at the Upper Border of the Pyriformis. These include the superior gluteal vessels and nerve.

When the gluteus maximus is reflected, the nerve and the vessels entering its deep surface, the *inferior gluteal*, can be traced to the lower border of the pyriformis. Next, at the lower border of the pyriformis the *sciatic nerve*, the largest nerve in the body, is identified and traced downward between the ischial tuberosity and the greater trochanter. Rarely, the nerve may emerge through the center of, or at the upper margin of, the pyriformis. Because its branches to the hamstrings spring from its medial side, the nerve may be safely dissected only from the lateral side.

Next, the superficial branches of the *superior gluteal vessels and nerve* are identified where they appear between the adjacent borders of the gluteal medius and the pyriformis. This can be done by following upward the posterior free border of the greater trochanter to the tendon of insertion of the gluteus medius. Then the lower border of the muscle is followed medially.

Superior Gluteal Nerve. This nerve runs with the deep branches of the superior gluteal vessels between the gluteus medius and minimus, supplying these muscles as well as the tensor fascia latae. These muscles act as abductors and external rotators of the thigh.

Inferior Gluteal Nerve. This emerges from the lower part of the greater sciatic foramen in company with the inferior gluteal vessels and the posterior cutaneous nerve of the thigh. The inferior gluteal vessels and nerve supply the gluteus maximus.

Gluteus Medius and Minimus. These fan-shaped and functionally similar muscles arise from most of the lateral surface of the ilium and insert into the lateral and anterior aspect of the greater trochanter. They function as abductors of the thigh. A bursa lies beneath their tendons of insertion over the anterior

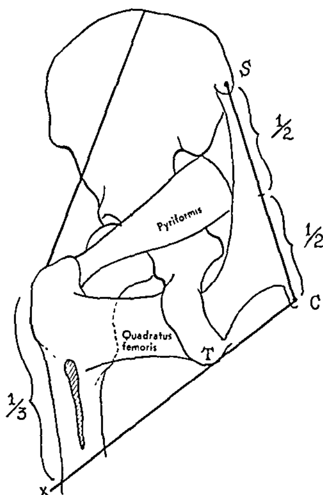


FIG 344. Outline of the gluteus maximus. This has often been described as a rhomboid whose upper and lower borders are parallel However, the origin of the upper border varies from several fingers to a full hand's breadth in front of the posterior superior iliac spine. The upper border extends to the tip of the greater trochanter. The lower border extends from the tip of the coccyx across the ischial tuberosity to the shaft of the femur at the junction of the upper third with the lower two thirds The midpoint of a line drawn from the posterior superior iliac spine to the tip of the coccyx designates the level of the lower border of the pyriformis. (Redrawn from Grant: A Method of Anatomy, ed. 4, Baltimore, Williams & Wilkins)

psoas fascia and appears as an abscess at the hip region. Tension within the psoas fascia produces a relative muscle shortening which clinically is apparent as a flexed hip Passive attempts at extension of the hip tense the muscle and cause pain.

The iliopsoas is the chief flexor of the thigh. Conversely, if the lower limbs are fixed, it is a flexor of the pelvis as when the trunk is raised from a recumbent position.

B. The Gluteal Region

Bony Landmarks. These include the *iliac crest*, the *anterior superior iliac spine*, the *posterior superior iliac spine* and the *ischial tuberosity*. The highest point of the crest corresponds to the level of the spinous process of the 4th lumbar vertebra. A line joining the posterior superior iliac spines crosses the second sacral segment which is the distal extremity of the dura. This line also crosses, when projected laterally, the middle of the sacro-iliac joint. A line drawn horizontally at the level of the ischial tuberosity crosses the lesser trochanter of the femur. This is a useful landmark in determining the level of osteotomy of the femur.

A line drawn from a point on the iliac crest a hand's breadth in front of the posterior superior spine directly outward to the tip of the greater trochanter represents the upper border of the gluteus maximus muscle. The lower border of this muscle lies along a line from the tip of the coccyx and extending through the ischial tuberosity.

By drawing a line from the posterior superior spine to the tip of the coccyx, the midpoint of this line extended out to the tip of the greater trochanter corresponds to the lower border of the pyriformis muscle.

Cutaneous Nerves. Branches of the posterior rami of the first 3 lumbar nerves cross the crest at the lateral border of the sacrospinalis. The lateral cutaneous branches of the anterior rami of the last thoracic and the first lumbar (iliohypogastric) nerves cross the crest anteriorly, one in front of the tubercle and the other behind it. Incisions made over the crest often sever these nerves, and painful neuromata develop. At the lower border of the maximus, several branches from the posterior cutaneous nerve of the thigh curve around the edge of the muscle and run upward

Gluteus Maximus. The deep fascia over this muscle is thin and transparent in contrast with a dense, opaque, pearly white fascia immediately in front of the maximus and overlying the gluteus medius. The gluteus maxi-

spine, is the origin of the sartorius muscle and the inguinal ligament. Below the anterior superior iliac spine, the anterior inferior iliac spine gives origin to the direct head of the rectus femoris. At the posterior extremity of the crest is the posterior superior iliac spine. Below this is the posterior inferior iliac spine which lies at the posterior extremity of the sacro-iliac joint. This is the site at which the sacro-iliac joint is most superficial and accessible for detecting tenderness. The posterior portion of the ilium forms a wing which partially overlies the sacrum, while its inner aspect articulates with the lateral irregular border of the sacrum. The ligaments surrounding the joint are quite strong and rarely disrupted. (See Sacro-iliac Strain in section on the Back.)

The outer aspect of the ilium gives attachment to the gluteal muscles. The gluteus medius occupies an extensive area below the iliac crest. In front, below and beneath the medius, a large area is occupied by the gluteus minimus. A small area remaining behind the medius is for the gluteus maximus and a portion of the sacrotuberous ligament. Between the gluteus minimus and the superior acetabular rim is a small area to which is attached the iliofemoral ligament and the reflected head of the rectus femoris.

The structure of the ilium is mainly cancellous and is particularly thick about the posterosuperior iliac spine, which provides an excellent source of bone transplants.

Ischium. The body of the ischium is inferior to the acetabulum and projects a *tuberosity* downward which gives origin to the hamstrings. A traction apophysis surmounts the tuberosity and may be avulsed by violent muscular contraction. The medial border of the tuberosity gives attachment to the sacrotuberous ligament above which the obturator internus passes laterally out of the lesser sciatic notch. From the lateral border of the tuberosity springs the quadratus femoris which passes laterally to the femur.

The tuberosity forms a palpable large prominence which supports the body when sitting. Between this prominence and that of the greater trochanter is a depression through which the sciatic nerve runs.

Sciatic Notches. The greater and the lesser

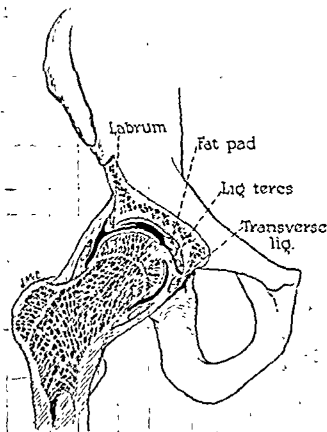


Fig. 345. Cross section of the normal hip. The position of the labrum, the fat pad, the ligamentum teres and the transverse ligament and the contour of the roof should especially be noted.

sciatic notches are located between the postero-inferior iliac spine and the ischial tuberosity. The *ischial spine* and its *sacrospinous ligament* separate the notches.

Sacrotuberous Ligament. This thick strong band extends from the postero-inferior iliac spine and the posterior aspect of the sacrum to the ischial tuberosity and gives origin to the gluteus maximus.

Ramus of the Ischium. This projects forward to join the inferior pubic ramus to form the inferior border of the *obturator foramen.*

Pubis. The pubis consists of a *body* lying medially, a *superior ascending ramus* and an *inferior descending ramus* which with the ischial ramus forms part of the *pubic arch.* The bodies of the pubic bones meet in the mid-line to form a syndesmosis, the *symphysis.* The outer surface of the body and the inferior ramus give attachment to the adductor muscles. At the lateral aspect of the body is the *pubic tubercle* to which attaches the

part of the lateral surface of the trochanter.

Tensor Fascia Latae. This arises from the anterior superior iliac spine and the adjacent portion of the outer lip of the iliac crest. It is enclosed between 2 layers of fascia. It is directed downward and slightly backward and is continued as the iliotibial tract. It is innervated by the superior gluteal nerve.

Quadratus Femoris. This oblong muscle appears to be the proximal portion of the adductor magnus. It arises from the lateral border of the ischial tuberosity and extends laterally to insert into the quadrate tubercle behind the greater trochanter and a linear area between this tubercle and the gluteal tuberosity near the lesser trochanter. The nerve to the quadratus lies beneath the sciatic nerve, which must be retracted before the nerve can be exposed. The nerve descends in front of the obturator internus and gemelli and enters the anterior aspect of the quadratus. This nerve supplies an articular branch and must be resected in performing denervation for hip pain of sciatic distribution.

Obturatorius Internus, Superior Gemellus and Inferior Gemellus. These muscles occupy the interval between the pyriformis and the quadratus. The obturatorius internus arises from the internal aspect of the innominate bone, makes a right-angled turn and goes out through the lesser sciatic foramen Its tendon passes across the posterior surface of the ischium and the capsule of the hip joint to reach the upper border of the greater trochanter. The superior gemellus arises from the ischial spine, and the inferior gemellus from the ischial tuberosity, both are inserted adjacent to the obturatorius internus

Superior Gluteal Arteries. These vessels hook around the margin of the greater sciatic foramen, supply muscles and provide a nutrient artery to the ilium. The inferior gluteal arteries send a branch to the sciatic nerve and cutaneous branches which accompany the posterior femoral cutaneous nerve of the thigh.

Posterior Femoral Cutaneous Nerve. This nerve is sensory First, it lies medial to the sciatic nerve, then posterior to it, and clings to the undersurface of the gluteus maximus. Its gluteal branch turns around the lower border of the gluteus maximus, and a perineal branch passes lateral to the ischial tuberosity, supplying the scrotum or the labium majus. The main nerve continues subfascially down the middle of the thigh and ends on the calf.

Iliotibial Tract. The fascia lata forms a thickened band on the lateral aspect of the thigh. This band is attached above to the tubercle and the anterior portion of the iliac crest and below to the lateral condyle of the tibia. Its upper portion is split into 2 layers which enclose the tensor fascia femoris It covers the greater trochanter, and below the trochanter receives the insertion of the gluteus maximus. Above the insertion of the gluteus maximus, the posterior border of the tract is continuous with the thick fascia covering the gluteus medius. Below the insertion of the gluteus maximus, its anterior and posterior borders are continuous with the fascia lata, and its deep surface sends a fibrous septum medially, the lateral intermuscular septum, which attaches to the lateral supracondylar ridge and the linea aspera and separates the quadriceps from the hamstrings.

C. The Bony Pelvis

The 3 main component parts—the ilium, the ischium and the pubis—meet in a cup-shaped concavity, the *acetabulum,* which receives the femoral head. Within the acetabulum, the junction between these components in the growing individual is occupied by the *triradiate cartilage,* which disappears as fusion takes place at about the 16th year.

Ilium. This large flat fan-shaped bone lies above the acetabulum. Its crest is subcutaneous and during the growth period is surmounted by an apophysis. Ossification of the apophysis takes place in a line extending along the entire length of the crest. Completion of this line of ossification coincides with termination of longitudinal growth. This sign is utilized in the management of scoliosis of the idiopathic type to signify that no further progression of the spinal curve can take place.

To the inner aspect of the crest attaches the abdominal muscles in front and the quadratus lumborum and the abdominal muscles behind. The outer border of the crest gives attachment to, from before backward, the tensor fascia latae, the gluteus medius and minimus and the gluteus maximus. The anterior extremity of the crest, the anterior superior iliac

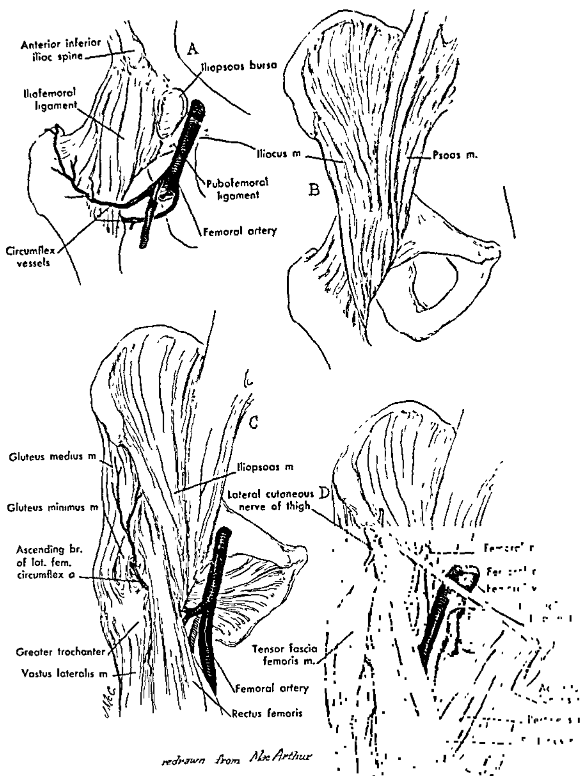


Fig. 348. Anatomy of the anterior aspect of the hip joint. The following points are worth noting: (A) The iliofemoral ligament of Bigelow (Y-ligament) is a strong supportive structure which attaches to the entire length of the intertrochanteric line. It tightens when the hip is fully extended. The lateral femoral circumflex vessels are shown encircling the base of the neck where penetrating branches pierce the capsule before passing along the neck toward the femoral head. Note the position of the iliopsoas bursa. It sometimes communicates with the joint cavity. (B) The iliopsoas muscle wraps itself around the capsule enroute to its insertion into the lesser trochanter. (C) This demonstrates the relative positions of the gluteus medius and minimus covering the hip above and laterally, and the rectus femoris and the vastus lateralis which clothe the hip and the upper femoral shaft anteriorly and laterally. (D) The superficial muscles, the tensor fascia latae and the sartorius are added, as are the femoral vessels and nerve. Note the point of exit of the lateral cutaneous nerve of the thigh. The interval between the sartorius and the tensor fascia latae is utilized in the iliofemoral approach.

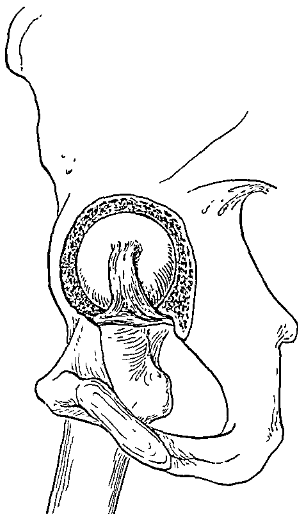


FIG 346. The normal hip visualized from the medial aspect with the acetabulum removed. Note that the acetabulum is deficient inferiorly where the notch is bridged by the transverse ligament.

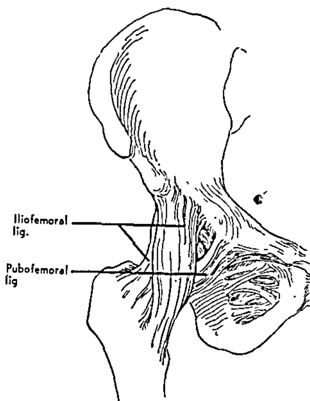


FIG. 347. Ligaments of the anterior aspect of the hip joint. The iliofemoral, or Y, ligament is the strong stabilizer of the hip in the extended position.

inguinal ligament Laterally, the superior ramus broadens to form one fifth of the acetabulum.

D. The Hip Joint

The hip joint is a ball-and-socket joint composed of the head of the femur and the acetabulum.

Ligaments. The capsule is attached proximally about the rim of the acetabulum. Distally, it is closely applied to the neck of the femur. Anteriorly, it attaches firmly to the intertrochanteric line. Posteriorly, it attaches weakly to the neck about ½ inch proximal to the intertrochanteric crest.

Three ligaments are represented by capsular thickenings. The *iliofemoral ligament,* shaped like an inverted Y is the thickest, strongest part of the capsule located anteriorly. As it passes distally toward the trochanteric line, it divides into 2 separate bands. The lowermost band passes obliquely downward and is tightened when the hip is fully extended. The Y-ligament is the chief stabilizer of the hip in the erect standing position It is ¼ inch thick and is rarely disrupted by trauma. Its preservation prevents excessive displacement and provides a fulcrum about which manipulative reduction of dislocation and fracture can be effected.

The *pubofemoral ligament* is a capsular thickening at the inferior aspect of the capsule. The *ischiofemoral ligament* is a weak band within the posterior capsule.

The *transverse ligament of the acetabulum* is a strong band of fibers which bridges and is attached to the margins of the acetabular notch It completes the rim of the acetabulum The vessels and the nerves enter the joint through the foramen beneath the ligament

The *labrum acetabulare* is a firm fibrocartilaginous ring which is fixed to the rim of the acetabulum and deepens the cavity.

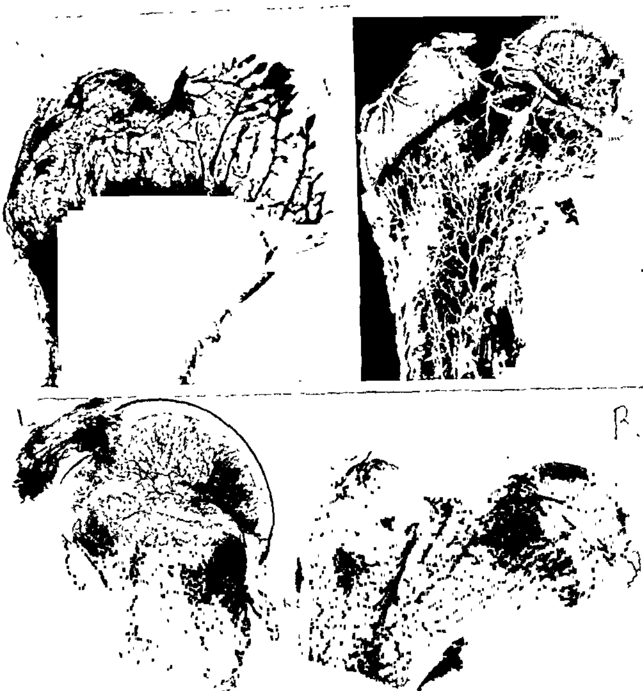


FIG. 349. The vascular patterns of the femoral head during growth. (*Top, left*) *At birth:* Main supply from lateral epiphyseal and metaphyseal vessels. (*Top, right*) *After 4 months:* Exclusive supply from lateral epiphyseal vessels. Cartilaginous growth plate is a barrier to metaphyseal vessels. (*Bottom, left*) *After age 7:* Vessels from the ligamentum teres penetrate and join the lateral epiphyseal vessels. Cartilaginous growth plate is still a barrier. Metaphysis is very vascular and becomes more so as epiphyseal fusion approaches. (*Bottom, right*) *Adulthood:* Epiphyseal plate barrier has disappeared. Anastomoses have formed between lateral epiphyseal, medial epiphyseal, superior metaphyseal and inferior metaphyseal vessels. (Trueta, J.: The normal vascular anatomy of the human femoral head during growth, J. Bone & Joint Surg. 39B:358)

epiphysis Severe manipulative efforts aimed at replacing a slipped epiphysis can destroy the lateral epiphyseal vessels and compromise circulation of the head, particularly when the ligamentum teres blood supply is inadequate. When reducing an old slipped epiphysis by open operation, it is advisable to perforate the epiphyseal plate to allow ready access of

The *ligamentum teres,* the ligament of the head of the femur, is flat and fan-shaped. Its narrow end is inserted into a pit in the femoral head; its flattened end is bifurcated and attached to the transverse ligament. A small artery runs along the ligamentum teres to the head of the femur. Before epiphyseal fusion, the artery of the ligamentum teres contributes to the blood supply to the epiphysis. Later, it is obliterated in most subjects.

Fat Pad. A mass of fibro-fatty tissue occupies the acetabular fossa. The articular vessels and nerve enter the fat pad.

Synovial Membrane. This membrane lines the inner surface of the capsule and covers the ligamentum teres and the labrum glenoidale. It is reflected distally upon the neck of the femur and covers the latter as far proximally as the margin of the articular cartilage.

At the distal synovial reflection, some of the fibers of the capsular ligament are likewise reflected and run upward on the femoral neck, raising the synovium as ridges called *retinacula.* These fibrous prolongations strengthen the external support and may prevent displacement after fracture.

Blood Supply. The arteries from the medial and the lateral femoral circumflex enter the capsule distally and posteriorly and pass proximally beneath the synovial membrane along the retinacula. They form the main source of blood supply to the femoral head. In fracture of the femoral neck, the central medullary vessels often prove to be inadequate, and aseptic necrosis of the head takes place.

Nerves. The nerves to the hip joint include (1) the nerve to the quadratus femoris, (2) the femoral through the nerve to the rectus femoris, (3) the anterior division of the obturator nerve, and (4) occasionally the accessory obturator. Denervation of the hip joint requires removal of all these nerves.

Vascular Anatomy of the Femoral Head[1,2]

The pathogenesis of many conditions about the hip, such as Legg-Perthes disease, slipped femoral epiphysis and osteoarthritis is unknown. Nevertheless, a basic consideration in investigation of the origin of these diseases is not only the extrinsic but also the intrinsic anatomy of the vascular supply. The following recently ascertained facts should be studied by the student as a possible springboard to further research into an understanding of these conditions.

Growth Period

At birth, nutrition of the femoral head is derived from 3 sources:

1. **Lateral epiphyseal** (from medial femoral circumflex). *These are a group of vessels* which enter the outer part of the femoral head in the region of the trochanteric notch and advance horizontally toward the center of the head.

2. **Metaphyseal** (from medial femoral circumflex). These are a series of straight vessels which ascend vertically through the cartilaginous head.

3. **Ligamentum teres vessels** (from acetabular branch of obturator). These barely supply a superficial portion of the femoral head.

Delay in ossification of the epiphysis so commonly observed in congenital dislocation could conceivably be explained by capsular stretching obliterating the lateral epiphyseal vessels while the metaphyseal vessels remain intact.

After 4 months of age, the ascending metaphyseal vessels decrease in size and number; they no longer pierce the cartilaginous plate as before, except for an occasional vessel ascending about the periphery. Therefore, *the main vascular supply at this time is through the lateral epiphyseal arteries.* Obstruction of these vessels could theoretically produce the picture of Legg-Perthes disease.

After the age of 7, the vessels from the ligamentum teres progressively penetrate more deeply and join the lateral epiphyseal vessels in supplying the head. The cartilaginous epiphyseal plate constitutes a barrier to metaphyseal blood flow until epiphyseal fusion takes place. Vascularity of the metaphysis is profuse during puberty in the period immediately preceding fusion, contributing greatly to weakening of the epiphysiometaphyseal junction and, consequently, slipping of the

[1] Trueta, J.: The normal vascular anatomy of the human femoral head during growth, J Bone & Joint Surg. 39B:358, 1957.

[2] Trueta, J., and Harrison, M. H. M. The normal vascular anatomy of the femoral head in the adult man, J. Bone & Joint Surg. 35B:442, 1953.

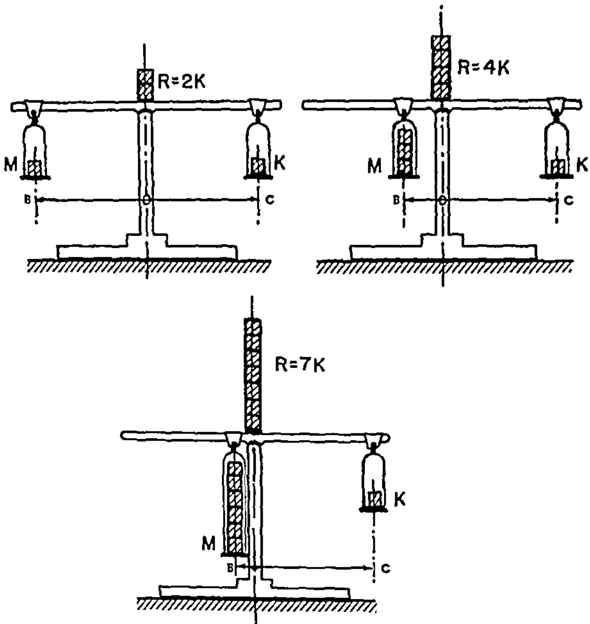


FIG. 351. Illustrating the forces operating upon the hip by utilizing the simple balance. These forces are represented by the vector R which is the sum of weights M and K. The force designated by M may represent the muscle force counterbalancing the body weight K.

(*Top, left*) In this diagram the superimposed weight R, which is equal to 2K, is divided equally at M and K, a perfectly balanced condition when the human body is standing erect.

(*Top, right*) The lever arm B to O is shortened to one third the length of lever arm O to C. In order to balance weight K, it is necessary for weight M to be 3 times the weight K to maintain equilibrium. This means that the center column supporting the balance beam must now withstand pressure 4 times that of K. This explains the excessive pressure upon the hip when the neck of the femur is shortened, or in marked valgus position

(*Bottom*) The short lever is one sixth that of the longer one. Therefore, the weight at M must be 6 times that at K and the forces exerted upon the central column will be the sum of M and K, or 7 times K. (Pauwels, F.: Spätfolgen der Schenkelhalsfraktur. *In* Bericht über unfallchirurgische Tagung am 12. und 13. Januar, 1952, *in* Stuttgart. Heft zur Unfallheilkunde, Heft zur Unfallheilkunde, Heft 45, Berlin, Springer)

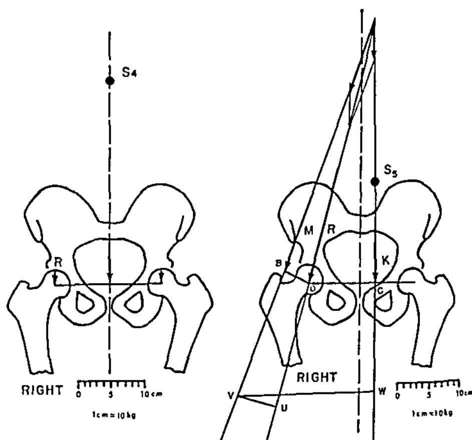


FIG. 350. (*Left*) When the patient stands, the weight of the body above the lower extremities, S_4, rests equally on the two normal hip joints. The static force on each hip, indicated by a short arrow, R, is one half the total superimposed weight. (*Right*) When the left lower extremity is lifted, e.g., during the swing phase of walking, the weight of the left lower extremity is added to the superimposed body weight, and moving the center of gravity to the left at S_5 where the total weight is represented by the heavy line, K. The abductors of the stationary right hip must pull downward on the right side of the pelvis at B (vector M) strongly enough to support and to maintain in equilibrium the weight K. The force downward on the fulcrum at O, the hip, is the sum of the vectors, M and K, expressed as R. The amounts of downward pull by the abductors at B and the downward force at C are directly related to the relative length of the levers B to O and O to C. If B to O is one third O to C, the downward pull of the abductor muscles at B, expressed as M, must be 3 times the downward force at C, which is K, if the forces are to balance. The total pressure on top of the femur, R, is the sum of M and K, or 4 times K. (Pauwels, F : Der Schenkelsbruch. Ein Mechanisches Problem, Stuttgart, Enke)

metaphyseal vessels into the head. After epiphyseal fusion, the adult sources of blood supply are brought together.

Adult Period

The *lateral epiphyseal arteries* anastomose with the *medial epiphyseal artery,* which enters at the fovea capitis. These cross the epiphysis horizontally and send branches mainly toward the articular margin. These branches pass through openings in the subchondral bone and reach the zone of calcified cartilage. Lesser branches are sent downward to anastomose with branches from the metaphyseal arteries.

Superior metaphyseal vessels enter the superior aspect of the femoral neck, and *inferior metaphyseal vessels* enter the inferior surface of the neck close to the articular cartilage. These metaphyseal arteries distribute small branches throughout the metaphysis as they ascend toward the epiphysis. Before entering the bone, the metaphyseal arteries form profuse anastomoses in the subsynovial tissues, the *circulus articuli vasculosus* of Hunter. This vascular network is deficient anteriorly over the neck. *The main blood supply to the femoral head is through the lateral epiphyseal arteries,* originating from the medial femoral circumflex.

It is important to note that the intrinsic vascular tree remains patent with advancing age. However, the arrangement of the vessels of smallest caliber is dependent upon distribution of the 2 types of marrow, yellow and red During growth years, red hemopoietic marrow

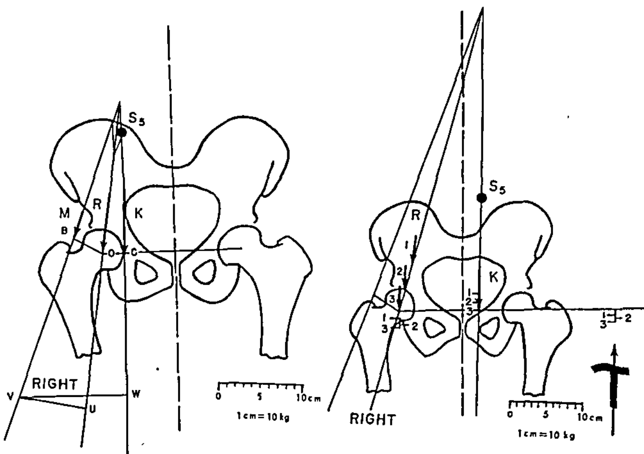


FIG. 353. When the trunk lists toward the side of the affected hip, the force of contraction required of the abductors is materially lessened and ambulation is improved. (*Left*) The trunk is deviated toward the right, shifting the center of gravity to the right (S_5). The lever arm, O to C, is reduced to less than the lever arm O to B. M becomes less than K, and the total force exerted upon the femoral head, R, is only slightly greater than the superimposed weight. (*Right*) The use of a supporting cane on the unaffected side reduces the body weight, K, and required abductor pull is lessened. The cane works through a long lever so that a moderate push upon the stick greatly relieves the pressure on the hip. The lines at the right labeled 1, 2 and 3 refer to the small amount of cane pressure which will reduce pressure upon the hip denoted by similar lines at the left. For example, 385 pounds pressure on the femoral head is reduced to 66 pounds pressure by exerting only 38 pounds of pressure upon the cane. (Pauwels, F.: Der Schenkelsbruch. Ein Mechanisches Problem, Stuttgart, Enke)

Clinical Applications

When a hip is in valgus, the short abductor lever arm requires tremendous abduction pull on the hip, and the resultant pressure on the head may be as much as 7 or 8 times the supported weight. To reduce the pressure and the pain the patient lists the trunk toward the hip and displaces the center of gravity in that direction. Consequently, less pull on the abductors is required, and force on the femoral head is reduced This is the characteristic waddle and limp of a coxa valga, a means of relieving stress on the hip. The secondary strain on the lumbar spine caused by this lateral lurching produces backache. Increased pressure on the femoral head increases degeneration. The use of a cane in the opposite hand by working through a long lever arm can reduce static force on the hip in multiples of pressure force exerted downward on the cane. Thus 20 pounds push on the stick can reduce static force on the opposite hip by 8 to 10 times that amount. When the femoral neck has been converted by osteotomy to a valgus position, the tremendous increase of load on the head makes it mandatory to relieve pressure by support at least until the head is strong enough The normal length of the femoral neck should be preserved wherever possible, particularly in prosthetic replace-

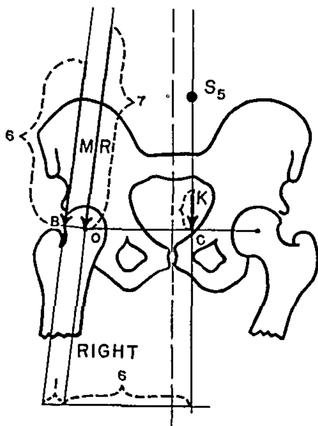


FIG. 352. The valgus hip, diagrammatic representation of forces. The pull of the abductors at B is at the end of an abnormally short lever arm, B to O, which is only one sixth of O to C. The pull downward at B must be 6 times K in order to maintain equilibrium when the left foot is raised. The total pressure upon the stationary right hip is 7 times K (Blount, W. P: J. Bone & Joint Surg 38A:695)

occupies most of the epiphysis and the metaphysis. In normal adult life, it is restricted to 2 areas, the metaphysis and a zone of epiphysis underlying the articular cartilage and the fovea capitis Most of the adult epiphysis is occupied by inactive yellow fat. Venous sinusoids, which are spacious thin-walled vessels, form profuse channels throughout the red marrow. In the yellow marrow, small blood vessels of capillary size are sparsely scattered between the fat

Normal articular cartilage is avascular.

MECHANICS OF THE HIP JOINT[3-7]

The hip joint is a ball-and-socket joint. In weight-bearing the pressure forces are transmitted to the head and the neck of the femur at an angle of 165° to 170° regardless of the position of the pelvis. The plane of force coincides with the strongly developed trabeculae which lie in the medial portion of the femoral neck and extend upward through the superomedial aspect of the femoral head. These trabeculae are in line with similar pressure trabeculae which start at the acetabulum and run upward and medial to the sacro-iliac joint The reacting force normally runs perpendicular to the cartilaginous epiphyseal plate.

When the weight of the body above the lower extremities rests equally on 2 normal hip joints, the static force on each hip is one half of the total, or less than one third of the total body weight (Fig. 350). When, for example, the left lower extremity is lifted as in the swing phase of walking, the weight of the left lower extremity is added to that of the body weight, and the center of body gravity, normally in the median sagittal plane, is displaced to the left (Fig. 351). The abductor muscles exert a counterbalancing force to maintain equilibrium. The pressure exerted on the head of the right femur is the sum of these 2 forces. Each force is related to the relative length of levers. If the abductor lever (B to O) is one third that of the lever arm from the head to the center of gravity (O to C), the downward pull of the abductors must be 3 times the force of gravity to maintain balance. Therefore, the total pressure on the head is 4 times the superimposed weight. The longer the abductor lever, (i.e., the more laterally placed insertion of the abductors), the less the ratio between the levers, the less the abduction force required to maintain balance, and the less the pressure force upon the femoral head.

[3] Blount, W. P. Presidential Address American Academy of Orthopedic Surgeons, January 30, 1956; J. Bone & Joint Surg. 38A·695, 1956

[4] Inman, V. T.. Functional aspects of the abductor muscles of the hip, J Bone & Joint Surg. 29 607, 1947.

[5] Osborne, G V, and Fahmi, W H: Oblique displacement osteotomy for osteoarthritis of the hip joint, J. Bone & Joint Surg. 32B 148, 1950

[6] Pauwels, F.: Der Schenkelsbruch. Ein Mechanisches Problem, Stuttgart, Enke, 1935.

[7] ———. Spatfolgen der Schenkelhalsfractur Bericht uber unfallchirurgische Tagung am 12 und 13 Januar, 1952, in Stuttgart Heft zur Unfallheilkunde, Heft 45, Berlin, Springer, 1953





FIG. 354. Pathology of osteoarthritis of the hip. Demonstrating the formation of bone and marrow within degenerate articular cartilage. Note that the articular cartilage covering that portion of the head exposed to maximum pressure is well preserved. The cartilage over the medial nonpressure area is degenerate. The trabeculae of the pressure system are prominent. The enclosed area blocked out in the illustration at the left is shown at the right under higher magnification. Bone and marrow have been formed within the degenerate articular cartilage. (Harrison, M. H. M., Schajowicz, F., and Trueta, J.: Osteoarthritis of the hip: a study of the nature and evolution of the disease, J. Bone & Joint Surg. 35B: 598)

acetabulum transmits pressure forces to the head over a large area located superiorly and peripherally. A dense system of longitudinally disposed trabeculae are formed in the head and the neck immediately beneath this pressure area, apparently an architectural arrangement to protect against the stresses imposed on the bone. Over the pressure area the cartilage is thickest and contains the largest amount of chondroitin. The central and inferior surface of the head is apposed to the central soft fat pad of the acetabulum, and the extreme outer margin of the head lies free. At these last two sites of least pressure cartilaginous degeneration occurs more rapidly, the process apparently being accelerated by inactivity and lack of pressure. The degenerate cartilage is shed into the joint cavity and is engulfed by the synovial membrane where it disintegrates and is absorbed. This causes a chronic reactive hyperemia in the synovium which in consequence becomes hyperplastic with formation of multiple layers of surface cells and development of bunches of villi. Progressive fibrosis thickens the synovium, the subsynovial tissue and the overlying capsule.[14] Adhesions obliterate synovial pockets. The process is most pronounced at the inferior region of the joint, supposedly because the cartilaginous debris descends by gravity into the inferior synovial recesses.

In the normal mechanism of standing the hip is stable in extension, slight abduction and slight internal rotation. This permits the opposite side of the pelvis to be elevated in taking the next forward step with the oppo-

[14] Key, J. A.: Experimental arthritis, J. Bone & Joint Surg. 11:705, 1929.

ment operations. Maintenance of an adequate abductor lever will lessen pressure and enable the prosthesis to withstand stresses for a longer period of time.

When abductor paralysis exists, equilibrium cannot be attained. The individual shifts laterally and displaces the center of gravity over the affected hip so that forces are minimal and vertical. During the growth period the epiphyseal plate tends to remain perpendicular to these forces and consequently shifts to a horizontal position. The result is a coxa valga deformity. A similar mechanism is operative in valgus associated with congenital dislocation of the hip.

In the subluxated hip the greater trochanter is closer to the fulcrum point. In addition, the force acts only along the upper border of the acetabulum instead of being distributed over the whole. Therefore, excessive pressure is exerted over a small localized area and degeneration is inevitable. An adduction osteotomy to create a varus deformity will produce these effects: (1) the trochanter will be displaced a greater distance from the pelvis, thereby reducing the load on the femoral head; and (2) the line of force will be made to act upon the center about a large acetabular area. Relief of pain and fatigue is accomplished.[8]

DEGENERATIVE JOINT DISEASE

Etiology

Physiologic degeneration of articular cartilage takes place with advancing age. Chemically, it is characterized by loss of chondroitin sulfate which is proportionate to loss of elasticity. In the hip joint the process is accelerated by the following factors:

1. **Incongruity of articular surfaces,** causing abnormal friction Examples include irregularities of the femoral head after healed Legg-Perthes disease and after fractures of the articular surfaces.

2. **Instability,** causing abnormal mechanical friction. Examples include subluxation which progressively erodes the upper acetabular rim, and dysplasia of the hip with its shallow acetabulum.

3. **Ischemia.** Complete interruption of the blood supply to the femoral head occurs with severe traumatic dislocations and fractures through the femoral neck. The closer the fracture approaches the head the more likely the loss of circulation followed by the appearance of aseptic necrosis. The factor of insidious ischemia forms the basis for operations to restore the blood supply, e.g., transposition of muscle flaps.[9] Its role is questionable. The head in degenerative arthritis is vascular. The triangular density seen in the upper part of the head is not aseptic necrosis but vascular dense bone. Finally, people with severe circulatory disturbance, such as Buerger's disease or advanced arteriosclerosis, do not show an increased incidence of osteoarthritis of the hip

4. **Concentration of Pressure Load.** Normally, pressure forces in weight-bearing are widely distributed over the femoral head. When deformity, such as coxa vara or anteversion, concentrates the pressure forces over a small area, that area is likely to break down.

5. **Direct injury** to the cartilage, such as by infection or a missile. A direct blow can damage the cartilage without necessarily producing an open wound.

6. **Constitutional Causes.** These are many and include mainly obesity, hypothyroidism, pituitary dysfunction and the menopause.

7. **Idiopathic Causes.** At least 50 per cent can be classified thus. Recent investigation has revealed a high incidence of anteversion of the femoral neck, which constitutes an unrecognized deformity of congenital dysplasia.[10]

Pathogenesis[11, 12, 13]

Normally, when the femoral head is in contact with the acetabulum during weight-bearing, the peripherally placed labrum of the

[8] Pauwels, F Hip conditions of mechanical origin and their treatment by adduction osteotomy, Rev. chir. orthop, 37.22, 1951

[9] Venable, C. S, and Stuck, W. G.: Muscle flap transplant for the relief of painful monoarticular arthritis of the hip, Ann Surg 123:641, 1946.

[10] Merle D'Aubigné, R., and Postel, M · Functional results of hip arthroplasty with the acrylic prosthesis, J. Bone & Joint Surg 36A·451, 1954.

[11] Lloyd-Roberts, G. C · The role of capsular changes in osteoarthritis of the hip joint, J Bone & Joint Surg 35B:627, 1953

[12] Harrison, M H. M, Schajowicz, F., and Trueta, J.. Osteoarthritis of the hip A study of the nature and evolution of the disease. J. Bone & Joint Surg 35B·598, 1953.

[13] Lloyd-Roberts, G C: Osteoarthritis of the hip A study of the clinical pathology. J. Bone & Joint Surg. 37B·8, 1955.





FIG. 354. Pathology of osteoarthritis of the hip. Demonstrating the formation of bone and marrow within degenerate articular cartilage. Note that the articular cartilage covering that portion of the head exposed to maximum pressure is well preserved. The cartilage over the medial nonpressure area is degenerate. The trabeculae of the pressure system are prominent. The enclosed area blocked out in the illustration at the left is shown at the right under higher magnification Bone and marrow have been formed within the degenerate articular cartilage. (Harrison, M. H. M., Schajowicz, F., and Trueta, J.: Osteoarthritis of the hip: a study of the nature and evolution of the disease, J. Bone & Joint Surg. 35B: 598)

acetabulum transmits pressure forces to the head over a large area located superiorly and peripherally. A dense system of longitudinally disposed trabeculae are formed in the head and the neck immediately beneath this pressure area, apparently an architectural arrangement to protect against the stresses imposed on the bone. Over the pressure area the cartilage is thickest and contains the largest amount of chondroitin. The central and inferior surface of the head is apposed to the central soft fat pad of the acetabulum, and the extreme outer margin of the head lies free. At these last two sites of least pressure cartilaginous degeneration occurs more rapidly, the process apparently being accelerated by inactivity and lack of pressure. The degenerate cartilage is shed into the joint cavity and is engulfed by the synovial membrane where it disintegrates and is absorbed. This causes a chronic reactive hyperemia in the synovium which in consequence becomes hyperplastic with formation of multiple layers of surface cells and development of bunches of villi. Progressive fibrosis thickens the synovium, the subsynovial tissue and the overlying capsule.[14] Adhesions obliterate synovial pockets. The process is most pronounced at the inferior region of the joint, supposedly because the cartilaginous debris descends by gravity into the inferior synovial recesses.

In the normal mechanism of standing the hip is stable in extension, slight abduction and slight internal rotation. This permits the opposite side of the pelvis to be elevated in taking the next forward step with the oppo-

[14] Key, J. A.: Experimental arthritis, J. Bone & Joint Surg 11:705, 1929.

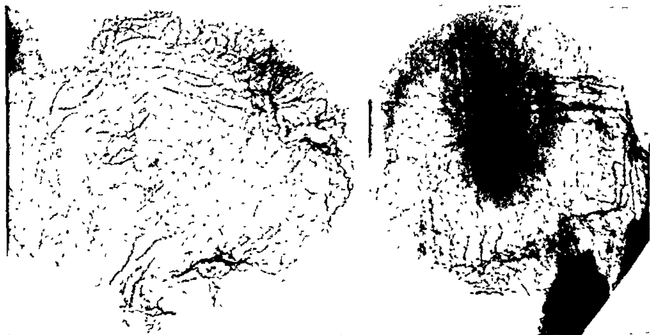


FIG. 355. Pathology of osteoarthritis of the hip. Angiograms of the femoral heads of a subject aged 76. The head at the left, the site of advanced osteoarthritis, displays an increased number of vessels which are dilated, especially at the weight-bearing area of the epiphysis. The head at the right, comparatively unaffected by osteoarthritis, shows a lesser degree of vascularity. (Harrison, M. H. M., Schajowicz, F., and Trueta, J.: Osteoarthritis of the hip: a study of the nature and evolution of the disease, J. Bone & Joint Surg. 35B:598)

site limb The posterior and inferior capsule is taut in this position. When fibrosis has caused thickening, shortening, and loss of elasticity of the inferior capsule, the femur is pulled into an opposite deformity of flexion, adduction and external rotation. Any attempt to attain the standing position imposes severe stretching of the sensitive capsule, causing



FIG. 356. Severe degenerative arthritis of the hip.

pain. The sensory nerve fibers to the inferior capsule arise from the obturator nerve, which supplies the adductors. These muscles undergo reflex spasm which accentuates the deformity. Persistence of spasm leads to progressive fibrosis and shortening of these muscles, particularly at their musculotendinous junctions, thereby perpetuating the deformity. The fibrotic small external rotator muscles may adhere to the posterior capsule.

Flexion, adduction and external rotation deformity narrow the area of weight-bearing pressure on the superior aspect of the femoral head. Therefore, the pressure forces are increased, and the degenerating cartilage succumbs and breaks down rapidly. In an effort to strengthen the area deprived of cartilage the process of new bone formation is accelerated. The deep layer of calcified cartilage thickens. The underlying medulla becomes increasingly vascular. Vessels penetrate the subchondral plate and lay down new bone superficial to the plate. Where external pressure is absent at the inferomedial aspect of the head and at the extreme outer margin,



FIG. 357. Demonstrating the effect of loose cartilage fragments as they are engulfed by the synovial membrane. (*Top*) Osteoarthritic hip. This field shows the typical histologic appearance of the cartilage fragments. The space between the main fragments probably contained a fragment which was dislodged during preparation (× 280). (*Bottom*) Synovial membrane of rabbit four weeks after injection of cartilage fragments. Note the generalized synovial hyperplasia and the fibroblastic response in the deeper layers (× 112). (Lloyd-Roberts, G. C.: Role of capsular changes in osteoarthritis of the hip joint, J. Bone & Joint Surg. 35B: 627-642)

ossification proceeds unopposed with the formation of large protruding osteophytes. Where the synovium attaches about the margin of the head the large synovial vessels penetrate the bone and are concerned chiefly with the formation of these marginal bony excrescences. Where pressure is greatest at the superolateral aspect, old and new bone is compressed, eroded, and suffers minute trabecular fractures. The concentration of compressed bone appears to be dense and eburnated, suggesting aseptic necrosis, but actually

this is dense vascular bone. Further destruction causes extrusion of more debris of degenerate cartilage and bone plus intra-articular bleeding, which serve to continue synovial and capsular hyperemia and fibrosis. A vicious cycle is established.



Pathology

The following are characteristic findings:

Articular Cartilage. This undergoes early degeneration, fibrillation, and fragmentation. Chondrocytes are lost or abnormal. The matrix loses its metachromatic staining quality. *Cracks extend through the entire cartilage to the bone.* The basal calcified layer thickens. Vascular channels pass from the marrow through the subchondral plate and form new bone superficial to the plate. Eventually, the cartilage is detached.

Synovial Membrane. This is congested and fibrous. Villous formation occurs in clusters. Surface cell layers are multiple. Hemosiderin may be present. Degenerate and disintegrating minute fragments of bone and cartilage lie just below the surface. In the deeper layers one occasionally sees well-staining new bone and cartilage metaplastic formations. Fibrosis is progressive, and intra-articular adhesions form. Pathologic changes are greatest below the neck of the femur.



Fig. 358. (*Top*) Section of cyst communicating with surface of femoral head. (× 110) (*Bottom*) Section showing subchondral plate and deep cartilage layers left behind by advancing osteophyte. (× 32) It appears that active osteogenesis is a reactive process designed to thicken the subchondral plate and is particularly pronounced at the periphery where it cannot be impeded by friction or pressure. Inset—Microradiograph illustrating position of section. (Lloyd-Roberts, G. C.: Osteoarthritis of the hip, J. Bone & Joint Surg. 37B:8-47)

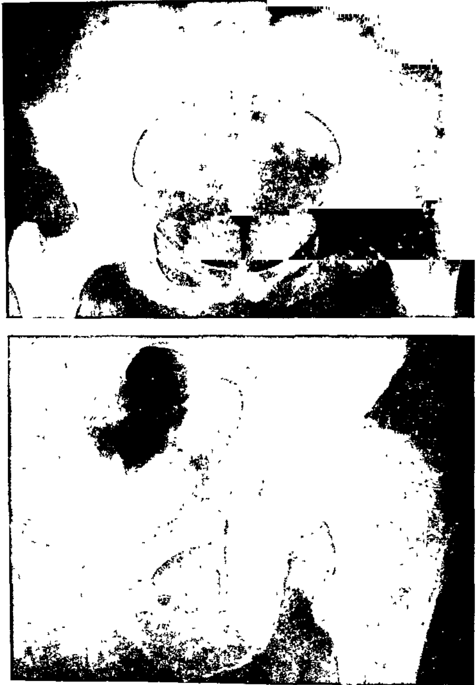


FIG. 359. (*Top*) Malum coxae senilis. The femoral head is sclerotic, flattened, and irregular in contour. The opposing articular surface of the acetabulum presents similar changes but to a lesser degree. The joint space is narrow, indicating loss of cartilage. Such a picture generally results from aseptic necrosis of the femoral head following fracture through the neck or dislocation. Often no history of trauma is obtainable. (*Bottom*) Showing sclerosis, flattening, and irregularity of the femoral head, narrowing of the joint space, and reactive degenerative changes in the opposite articular surface.

Subchondral Bone. This displays increased vascularity and new bone formation. The cancellous bone is replaced by dense vascular bone, particularly at the site of maximum cartilage damage. This bone is less resistant to pressure and friction than cartilage. It readily wears away, causing flattening of the head and producing bone debris.

Cysts lie within the area of subchondral bone beneath the point of maximum cartilage damage. Each cyst contains vascular fibrous tissue, is surrounded by dense bone and communicates through a channel with the joint. Cysts heal when excessive pressure is removed or redistributed, as for example after osteotomy.

Bony Ridge on the Femoral Neck. When the disease is well advanced and the capsule is shortened, a bony overgrowth appears on the undersurface of the femoral neck. It may be due to traction imposed on the periosteum by the capsule.

Osteophytes occur anywhere except in the upper quadrant. They arise by extension of newly formed bone into degenerate articular cartilage, where no friction or pressure can

retard its growth. It is not concerned with pain and rarely is the source of loose joint mice. Rather, the osteocartilaginous bodies commonly found arise by synovial metaplasia and extrusion.

Capsule. Fibrosis and hyperemia are the main findings. The inferior and posterior portions are chiefly affected. Occasionally, amorphous calcium salts are deposited in the superior capsule.

Clinical Picture

Course. At first, repeated attacks of slight pain lasting only a day or two are experienced An attack may be initiated by prolonged weight-bearing activity or occasionally by trauma, such as a misstep or a twisting strain. A sensation of stiffness about the hip appears after rest, and freer movement is obtained with activity. A protective limp due to muscle spasm is present. The pain becomes progressively worse in degree and duration, stiffness and restriction of hip motion are more persistent, and limp is permanent because of associated deformity. Eventually, the hip is stiffened, and the pain is lessened, because of severe restriction of motion.

Pain is located about the hip anteriorly, laterally, or posteriorly, depending on the site of acute inflammation, and is referred along the anterior and medial aspect of the thigh toward the inner aspect of the knee. It is accentuated by weight-bearing and movement in abduction, internal rotation and extension. Cold and humid weather intensifies the discomfort. Rest, heat and salicylates reduce the pain.

Tenderness is located over the site of capsular inflammation. The hip is most accessible to palpation posteriorly between the greater trochanter and the tuber ischium

Muscle spasm develops during an acute inflammatory incident and chiefly affects the adductors.

Deformity. Flexion, adduction and external rotation at first are due to spasm and later are rendered permanent by capsular contracture. The adduction contracture causes an apparent shortening. This in turn causes secondary lateral deviation of the lumbar spine. Hip flexion contracture, identified by the Thomas test, causes pelvic obliquity and secondary accentuation of lumbar lordosis. Lumbosacral pain is a consequence of hip contracture.

Fabere Test. The term "Fabere" is derived from the initial letters of "flexion," "abduction" and "external rotation," identifying the passive movements of the hip while eliciting the test. The foot of the affected extremity is placed upon the opposite knee, and the ipsolateral knee is forced outward and backward toward the table. The abduction and external rotation maneuver strains the anterior and inferior capsular ligaments. A painful response indicates inflammatory hip disease.

Limitation of Motion. At first the hip is limited by capsular contraction in extension, abduction and external rotation. Later destruction of joint surfaces restricts motion in all directions.

Roentgenologic Findings

Early films are negative. Irregular subchondral sclerosis gradually appears. The joint space slowly narrows. Flattening of the head occurs at the superior pole. Beneath this an area of density develops in the shape of an inverted triangle, and cysts are contained within this area. Marginal bony excrescences appear particularly at the lower junction of the head and the neck where exostosis may be of large proportion. Excess bone forms adjacent to the inferior margin of the neck coincident with capsular contracture.

Treatment

Principles. Severe pain and limitation of motion may occur early without severe joint changes. Conservative treatment frequently can eliminate symptoms and restore normal function. Where severe bone changes foretell repeated attacks and progressive disability, nonsurgical treatment can often surprisingly minimize disability and prevent recurrent acute attacks. Only when pain and deformity are persistent, particularly in the young patient, is more energetic treatment indicated. Where prolonged walking and standing are prime considerations, an arthrodesis provides an immobile but painless and stable hip. Its disadvantages are prolonged immobilization postoperatively, difficulty in sitting comfortably, and the strain placed particularly on an

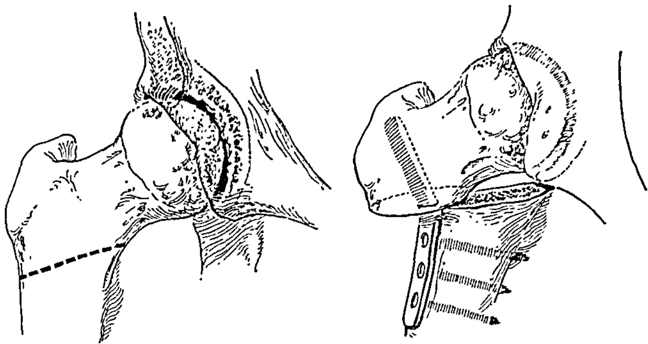


FIG. 360. The displacement osteotomy of McMurray. Insertion of a blade plate ensures fixation and permits early ambulation. Weight is borne on the distal fragment rather than on the hip joint.

arthritic lumbar spine. Therefore, it is contraindicated in bilateral hip disease and in the aged decrepit patient. A pertrochanteric or subtrochanteric osteotomy with medial displacement of the distal fragment provides stability by moving the weight-bearing line nearer to the mid-line of the body. At the same time the proximal fragment is rotated so that capsular tension is relieved and a less-affected surface of the head is approximated to the acetabulum; pain is relieved. Arthroplasty is used to preserve motion, especially when hip disease is bilateral or when a low-back condition is present. An inert metallic cup is interposed between the head and the acetabulum. Prosthetic replacement is seldom indicated. Failures are frequent enough to make subsequent procedures, because of loss of bone substance, very difficult problems.

Conservative Treatment. This consists of rest and abstinence from weight-bearing. Traction in abduction and application of heat and massage helps to overcome muscle spasm and prevent capsular contracture. The use of crutches or an ischial bearing caliper reduces weight-bearing pressure. Manipulation is restricted to gentle stretching of the inferior and anterior joint capsule. Injections of compound F are difficult to perform. Locating the joint interval with the needle is highly inaccurate, and the joint surfaces may be severely traumatized. When successful, the response is dramatic, but treatment must be repeated at intervals.

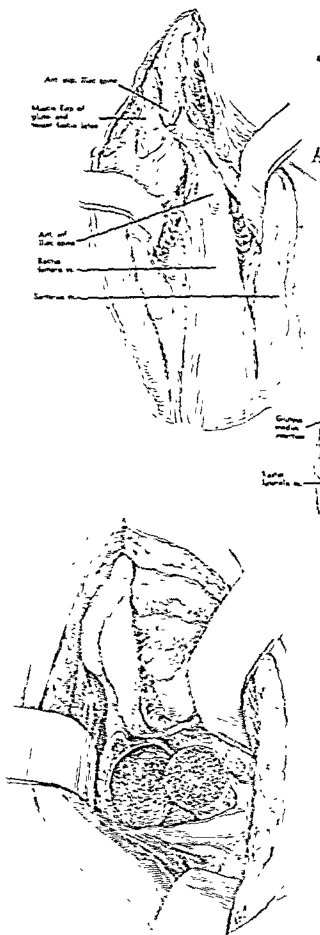
Oblique Displacement Osteotomy

The McMurray osteotomy consists of dividing the upper end of the femur so that the line of division passes from just below the greater trochanter upward and inward to a point above the lesser trochanter.[15] This permits medial displacement of the upper end of the shaft until it lies beneath the lower border of the hypertrophied acetabular rim where it receives the body weight. As a result, the load carried by the head of the femur is diminished, the proximal fragment may be slightly adducted, thereby relaxing the capsule and apposing a fresh articular surface, and pain is eliminated. Pressure within the hip joint is high when the femur bears weight in the typical adducted position, whereas such pressure is minimal or absent after displacement osteotomy.[16]

[15] McMurray, T. P.: Practice of Orthopedic Surgery, Baltimore, Williams & Wilkins, 1949.

[16] Osborne, G. V., and Fahrni, W. H: Oblique displacement osteotomy for osteoarthritis of the hip joint, J. Bone & Joint Surg 32B:148, 1950.

Fig. 361. Anterolateral exposure of the hip joint. In A, the origin of the sartorius is severed and reflected medially with muscles stripped from the medial surface of the ilium. Muscles are elevated from the lateral iliac surface, and wide separation of the 2 muscle units exposes the origin of the rectus femoris. The latter is severed and elevated, as in B, exposing the anterior capsule.



McMurray stipulated definite prerequisites before the procedure is used: (1) Flexion deformity must first be corrected, usually by traction; otherwise, the distal fragment may become displaced anteriorly with resultant nonunion. (2) There must be sufficient movement in the hip joint to allow adduction of the upper fragment. On the other hand, a fully *mobile hip will encourage* the proximal fragment to become abducted with loss of apposition and nonunion. Postoperatively, a hip spica for 3 or 4 months is necessary to ensure

Fig. 362. Exposure of the hip joint for cup arthroplasty. Sufficient bone is removed from both articular surfaces to ensure a loose fit. Although the capsule is well shown, it must be completely excised.

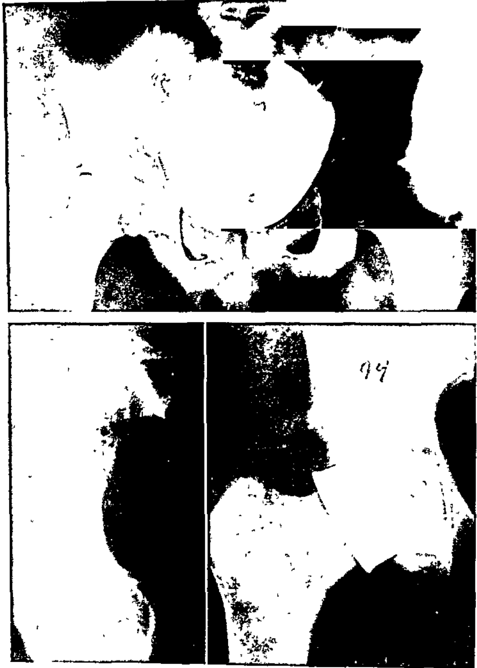


FIG. 363. Aseptic necrosis of the femoral head. The painful stiff hip was treated by cup arthroplasty.

union. A stiff knee is a common sequel of such prolonged immobilization. The use of a blade plate[17] will overcome all these handicaps Motion at the hip and the knee continues immediately after surgery, within a few weeks weight-bearing is permitted, and union occurs almost invariably.

Operative Technic. Through a lateral longitudinal incision the vastus lateralis is severed at its origin from the greater trochanter and cut along its posterior attachment and elevated forward and medially, exposing the lateral aspect of the femoral shaft. The osteotome is started just below the base of the greater trochanter and directed upward and inward at an angle of 40° to emerge medially between the lesser trochanter and the head of the femur. The proximal end of the distal fragment is displaced medially under the cotyloid ligament of the hip joint by direct medial pressure against the upper shaft and by wide abduction of the distal limb. A straight blade plate is inserted from below into the cancellous bone of the upper fragment. The distal half of the plate should

[17] Blount, W. P Blade-plate internal fixation for high femoral osteotomies, J. Bone & Joint Surg 25:319, 1943.



Fig. 364. Vitallium cup arthroplasty performed for painful degenerative arthritis following Legg-Calvé-Perthes disease. (Dr. William Schnute's case)

be bent outward at an angle of 140° so that when approximated and fixed by screws to the lateral aspect of the shaft the proximal fragment will be slightly adducted. A slight degree of flexion, abduction and neutral rotation of the shaft is advisable. The vastus origin is restored, and the wound is closed. Postoperatively, active and passive motion are started immediately. Weight-bearing is permitted within a few weeks. Pain should be reduced substantially, and good stability effected. The patient walks with the short "osteotomy stride." Failure to eradicate pain suggests inadequate displacement.

Cup Arthroplasty

The purpose in the use of cup arthroplasty is preservation of hip motion by reconstruction of the femoral head and acetabular surfaces and interposing a barrier substance.

Technic.[18] The skin incision extends along the iliac crest from the junction of the middle and the anterior thirds to the anterior superior iliac spine, then turns distally for several inches. The soft tissues (tensor fasciae latae and gluteus medius) are elevated subperiosteally from the lateral surface of the ilium.

[18] Smith-Petersen, M N.. Approach to and exposure of the hip joint for mold arthroplasty, J. Bone & Joint Surg. 31A:40, 1949

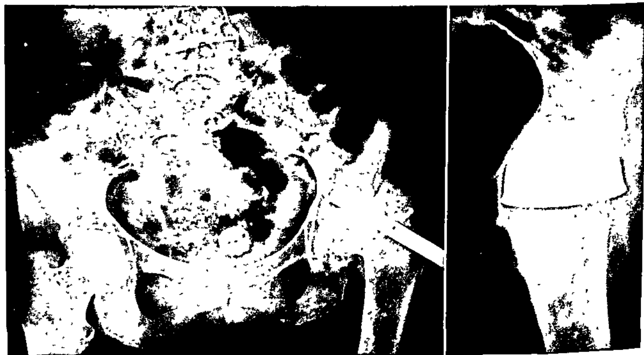


Fig. 365. Avascular necrosis of femoral head after fracture of neck Colonna reconstruction operation with cup arthroplasty. (Coventry, M. B.: Proc. Staff Meet. Mayo Clin. 29:48)

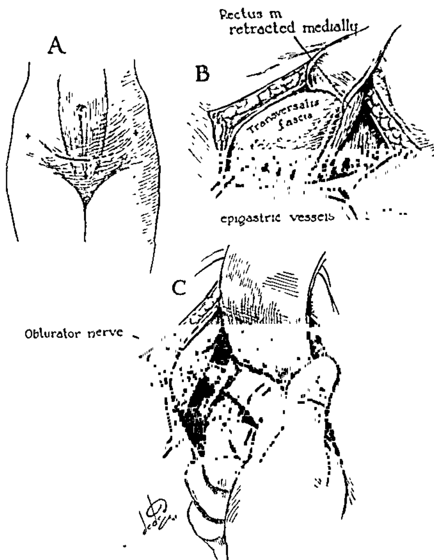


FIG. 366. Intrapelvic technic of obturator neurotomy. (Obletz, B. E., *et al.*: Early effects of partial sensory denervation of the hip for relief of pain in chronic arthritis, J. Bone & Joint Surg. 31A:805-814)

The femoral fascia between the tensor fasciae and the sartorius is incised, exposing a fat compartment covering the rectus femoris and extending medially behind the sartorius to cover the iliopsoas. This compartment is penetrated by blunt dissection. The anterior capsule is exposed, and the deep iliac fascia is incised between the main body of the iliacus muscle and the small lateral portion originating from the anterior inferior iliac spine. Care is taken to preserve the motor fibers of the femoral nerve to the rectus which lie on the anterior surface of the iliacus.

Between the inferior capsule and iliopsoas tendon is a fat pad through which run articular vessels from the lateral femoral circumflex artery. This area is developed by retraction of the muscle medially to expose the inferior aspect of the capsule adequately.

The origins of the rectus and the iliopsoas are severed close to the anterior inferior iliac spine and are reflected downward. Next, the antero-inferior iliac spine and the anterior acetabular wall are osteotomized, and the loose bone structure is excised with all of the fibrous capsule and synovial membrane. The distal capsular attachment must be avoided, since the circulation to the head and the neck enters at that point. The hip is dislocated. Both articular surfaces are denuded of cartilage and bone, and the contours are reshaped so that a large deep acetabular cavity results. Exposed cancellous bone surfaces bleed freely and have a tendency to proliferate and create spurs. Diathermy cauterization diminishes this tendency. Deepening of the acetabulum is done concentrically with suitable gouges, and final smoothing is accomplished with a reamer.

A deep cup is placed over the head, and the hip is reduced. Freedom of motion and stability are tested before closure. An excessively loose cup will result in eccentric motion,

FIG. 367. Posterior denervation of the hip by section of the nerve to the quadratus femoris. (Obletz, B. E., *et al.:* Early effects of partial sensory denervation of the hip for relief of pain in chronic arthritis, J. Bone & Joint Surg. 31A:805-814)

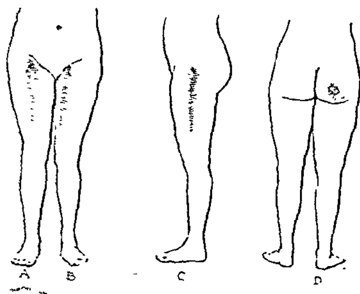
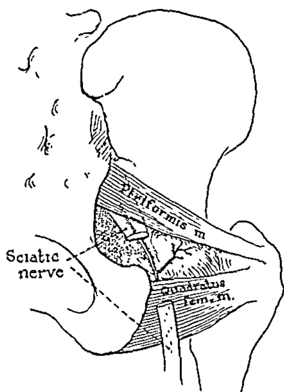
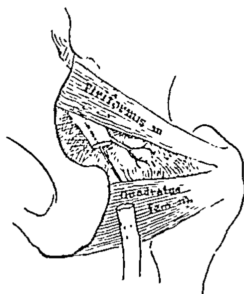


FIG. 368. Common pain patterns in chronic arthritis of the hip. A: Femoral pattern (anterior). B: Obturator pattern (medial). C: Lateral pattern. D: Posterior pattern. (Obletz, B. E., *et al.:* Early effects of partial sensory denervation of the hip for relief of pain in chronic arthritis, J. Bone & Joint Surg. 31A: 805-814)





wearing away of the head, and instability. It may be necessary, in addition, to perform a subcutaneous adductor tenotomy and displace the greater trochanter distally. Finally, the rectus and the sartorius origins are restored, and the tensor fasciae latae and the gluteus medius are sutured to the external oblique muscle. Postoperatively, only slight assistive exercises are permitted during the first 3 weeks while the muscle origins are healing.

FIG. 369. Posterior sensory nerve supply to the hip joint. Common pattern is shown at left. The sensory branch arises from the nerve to the quadratus femoris, which is given off by the sciatic nerve. Anatomic variation is seen at right. The sensory nerve arises as an independent nerve from the sciatic trunk in 5 of 24 dissections performed by Kaiser. (Obletz, B. E., *et al.:* Early effects of partial sensory denervation of the hip for relief of pain in chronic arthritis, J. Bone & Joint Surg. 31A: 805-814)

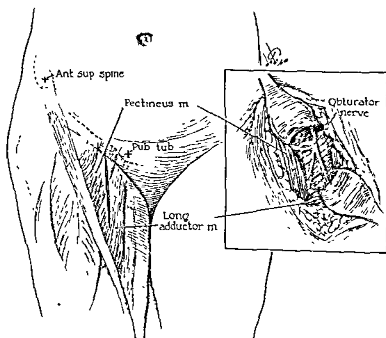


FIG. 370. Extrapelvic technic of obturator neurotomy. (Obletz, B. E., *et al.:* Early effects of partial sensory denervation of the hip for relief of pain in chronic arthritis, J. Bone & Joint Surg. 31A:805-814)

A
B
Gluteus max split & retracted
Piriformis m
Sciatic nerve
Reflected short rotators protecting sciatic nerve
C
Quadratus femoris m
to quadratus femoris m
uadratus femoris m

Fig. 367. Posterior denervation of the hip by section of the nerve to the quadratus femoris (Obletz, B. E., *et al.:* Early effects of partial sensory denervation of the hip for relief of pain in chronic arthritis, J. Bone & Joint Surg. 31A:805-814)

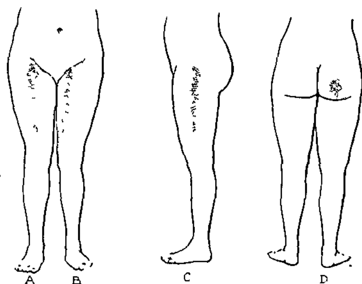


Fig. 368. Common pain patterns in chronic arthritis of the hip. A: Femoral pattern (anterior). B: Obturator pattern (medial). C: Lateral pattern. D: Posterior pattern. (Obletz, B. E., *et al.:* Early effects of partial sensory denervation of the hip for relief of pain in chronic arthritis, J. Bone & Joint Surg. 31A: 805-814)

peritoneum is freed and reflected posteriorly. The finger follows the lateral wall of the pelvis to the obturator foramen where the nerve is palpated. It is freed and pinched to note its innervation by spasm of the adductors. As much nerve as possible is removed to avoid possible regeneration. The epigastric vessels must be protected during the operation. The wound is closed in layers.

Extrapelvic Obturator Neurectomy. A transverse incision is made just below the inguinal ligament over the anteromedial aspect of the thigh. The pectineus and the adductor longus muscles are exposed and separated. In the interval is found the obturator nerve. The anterior branch lies between the adductor longus and the subjacent brevis. It is traced backward to the obturator foramen where the main nerve is found to divide into anterior and posterior branches. At least 1 inch of both branches is removed. Lateral branches, when found, are likewise excised.

Posterior Denervation. The incision is made from the posterosuperior iliac spine to the greater trochanter. The gluteus maximus is split near the trochanter, and the subgluteal fat in which the sciatic nerve is embedded is exposed. The nerve is protected, and the fat is divided to expose the underlying quadratus femoris, inferior gemellus, obturator internus, superior gemellus and pyriformis muscles. A blunt instrument is placed under the gemelli and the obturator internus, which are severed close to their insertion and retracted medially. The nerve to the quadratus runs perpendicular to the muscle and enters its undersurface near the ischium. The nerve lies directly on the bone and is covered by a very dense fibrous fascia. The nerve is freed and removed as high as possible. The nerve to the inferior gemellus, which also gives rise to articular branches, should be sought for and removed. Occasionally, a large sensory branch comes off the sciatic nerve just above the nerve to the quadratus and runs parallel with this nerve as far as the hip joint where it enters the hip capsule. If found, it too must be resected. The rotator muscles are restored to the greater trochanter, and the skin is closed.

Results. Adductor spasm is relieved, and sensation is diminished over the medial aspect of the thigh. Weakness in hip flexion is due to loss of the reinforcing action of the adductors.



Fig. 372. Hip prosthesis, Judet type. Roentgenographic appearance.

Such patients are unable to cross their legs; they lack endurance in walking, and sometimes even lack stability. It is preferable to combine obturator neurectomy with a cup arthroplasty.

Arthrodesis of the Hip

Fusion of the hip is the only certain method for eliminating pain and providing stability. If the hip has gradually stiffened by degeneration over several years, the spine at the same time compensates by increased mobility. By this mechanism sitting and bending are accomplished. On the other hand, if a mobile hip is suddenly deprived of its motion, the spine cannot adjust immediately, and backache results. Therefore, in contemplating arthrodesis of a movable hip it is wise to postpone surgery and provide the patient with a weight-bearing aid for a time until further compensation takes place. Further postoperative backache is less probable if all hip deformity is corrected and fixation in a neutral position is accomplished.

The operation is a formidable one and requires prolonged immobilization which may cause a stiff knee, a severe disability when the hip is also stiff. Therefore, it is better to treat aged and decrepit patients by the McMurray osteotomy with blade-plate fixation, or by cup arthroplasty.

Surgical Procedure. Any method of arthrodesis described in the section on Tuberculosis of the Hip may be used. In addition, internal

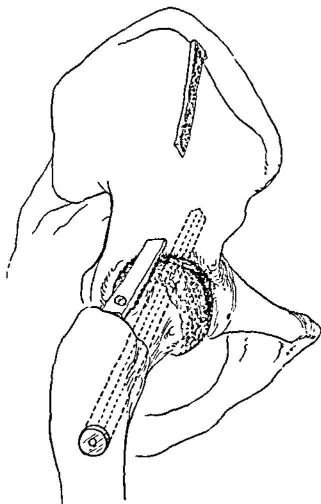


FIG. 371. Watson-Jones arthrodesis of the hip. The head and the acetabulum are thoroughly denuded. The nail is inserted through the neck and the head so that 1 inch of the nail engages in the pelvis. A bone graft removed from the ilium is inserted through a slot in the superior acetabular rim and fixed by a screw to the neck (Watson-Jones, Sir R., and Robinson, W. C.: Arthrodesis of the osteoarthritic joint, J. Bone & Joint Surg. 38B:353)

Thereafter, active exercises are instituted, with particular attention directed to the abductors and the extensors.

Denervation of the Hip

Pain is the major disabling factor in chronic arthritis of the hip. Theoretically, removal of the nerve supply to the capsule should abolish pain. However, in practice complete relief of pain is rarely obtained, but satisfactory improvement occurs in most cases The advantage of the operation lies in its simplicity and rapid convalescence. At the present time the procedure is chiefly confined to obturator neurectomy to overcome adduction deformity preliminary to arthroplasty, or as an independent operation in spastic paralysis. Denervations may be done when standard reconstructive procedures are contraindicated.[19,20]

Patterns of Pain About the Hip

1. *Obturator Pattern.* Pain is deep in the groin and radiates along the medial thigh to the knee.

2. *Posterior Pattern.* Pain is in the buttock behind the hip joint.

3. *Femoral Pattern.* Pain radiates from the front of the hip joint distally along the anterior thigh.

4. *Lateral Pattern.* Pain over the great trochanter radiates along the lateral thigh to the knee.

The obturator pattern is seen in nearly all patients and often is combined with the posterior pattern. Other types are rare except when associated with the obturator pattern.

Nerve Supply to the Hip Joint

1. *Obturator nerve* from ventral divisions of L2, 3, 4. An *accessory obturator nerve* is present in about 25 per cent of patients; from ventral divisions of L3, 4.

2. *Femoral nerve* from dorsal divisions of L2, 3, 4, by way of branches to the rectus femoris.

3. *Sciatic nerve* from ventral divisions of L5 and S1, by way of the branch to the quadratus femoris.

The muscles supplied by these nerves undergo spasm and contracture and produce the characteristic flexion, adduction and external rotation deformity.

Surgical Technic. Obturator nerve section and cutting the nerve to the quadratus femoris is the simplest and least harmful procedure

Intrapelvic Obturator Neurectomy.[21] The bladder is kept decompressed by an indwelling catheter, and the patient is placed in the Trendelenburg position. A Pfannenstiel incision is made, and the lower rectus sheath is split transversely. The rectus is retracted medially, and by blunt dissection the lower

[19] Kaiser, R. A · Obturator neurectomy for coxalgia, J Bone & Joint Surg. 31A:815, 1949.

[20] Obletz, B E, *et al* Chronic arthritis of hip effects of partial denervation, J. Bone & Joint Surg 31A 805, 1949

[21] Chandler, F A, and Seidler, F.: Intrapelvic extraperitoneal resection of the obturator nerve, Surg, Gynec. & Obst 69:101, 1939.

Indications. The most common indications are a stiff, deformed and painful *osteoarthritic joint, nonunion* of the femoral neck, *aseptic necrosis* of the head and *fresh fractures* of the neck in aged who cannot meet the restrictions imposed by hip nailing. Subcapital fractures are particularly liable to produce aseptic necrosis of the head and therefore are most suitable for prosthetic replacement. This fracture is noted for delay in healing, and the prolonged immobilization is dangerous to the aged patient. A prosthesis quickly restores activity. Conditions which cause *extensive destruction of the head and the neck*, e g., radium necrosis fractures after radium applications for malignancy of the cervix, should be treated by a prosthesis. When *hip fusion is contraindicated* by lumbar spine disease, prosthetic replacement is an alternative.

Technic of Insertion of Intramedullary Prosthesis.[26] Preoperatively, the size of the femoral head and the length of the femoral neck are determined by roentgenographic study of the opposite hip. At the time of surgery, various sizes of prostheses should be available.

The posterior approach to the hip joint is advisable, because bleeding is minimal, the gluteal muscles so necessary to hip stability are undisturbed, and the joint is exposed easily and widely. The patient lies on the side with the affected hip uppermost. The incision is made along the posterolateral aspect of the hip from the base of the greater trochanter and extends distally for several inches. Then the proximal end of the incision is extended for about 3 or 4 inches proximomedially along the lower third of the gluteus maximus and in line with its fibers

The aponeurotic insertion of the gluteus maximus into the iliotibial tract is severed, and the opening is extended through a muscle-splitting approach in the lower end of the muscle. The hip is flexed to 90°. The gluteus maximus is easily retracted upward so that the sciatic nerve can be identified and retracted and the small external rotator muscles brought into view. The branch of the sciatic to the quadratus should be sought and severed, as this may help to control postoperative pain. Fatty tissue behind the lower portion of the capsule of the hip joint is removed.

The insertions of the short external rotators are severed and retracted medially, the capsule coming into view. Ordinarily, it is sufficient only to sever the tendons of the obturator internus and the gemelli and occasionally the proximal portion of the quadratus. Then the capsule is incised along the axis of the neck and cut transversely at its distal attachment and reflected, exposing the base of the neck. The lesser trochanter is identified. The femoral head is dislocated by flexing the hip to a right angle, adducting the thigh and internally rotating the thigh by leverage on the leg while the knee is flexed to a right angle. At the conclusion of the maneuver, the foot should be faced toward the ceiling. If dislocation does not occur with ease, the neck is transected with a saw or an osteotome, the distal fragment is dislocated out of the wound, and the head is removed. Then the neck of the femur is cut across about ½ to ¾ inch above the lesser trochanter, in a plane preserving a normal anteversion of 20°. Osteophytes may be removed from the acetabular rim but rarely is it necessary to deepen the socket.

A special rasp corresponding to the size and the contour of the prosthetic stem is used to remove bone gradually from the medullary cavity. Before the prosthesis is inserted, small pieces of cancellous bone are wedged into the fenestrations of the stem. This encourages growth of bone through the openings and aids in locking the stem in place. Calipers measure the diameter of the acetabulum, and the proper prosthesis is inserted. The stem should fit snugly in the femoral shaft, often requiring a few blows on the inserter before the prosthesis finally settles into place. Reduction into the acetabulum is effected preferably by fully extending and applying traction to the extremity. The external rotator tendons are restored to their insertions, and the wound is closed loosely.

Postoperatively, the extremity is kept in extension and abduction for about 2 or 3 weeks as a precaution against dislocation. Gradually increased and guarded movement in bed is permitted and followed by short periods of graduated weight-bearing. A cane is often

[26] Moore, A. T.: The self-locking metal hip prosthesis, J. Bone & Joint Surg. 39A:811, 1957.



FIG. 373. Intramedullary prosthesis, Moore type. (Dr. Edward Compere's case)

fixation, which was contraindicated in infectious cases, may be used to ensure success of fusion. The contraindications are a stiff knee on the same side and a stiff hip on the opposite side.

Watson-Jones Arthrodesis [22] The hip is exposed through a Smith-Petersen incision, and the capsule is excised. The hip is dislocated anteriorly, if necessary by removal of the anterior lip of the acetabulum. All articular cartilage is removed. The hip is reduced, the limbs are placed parallel, and the affected limb is pulled down until it is level with the opposite limb. This step provides exactly the proper amount of abduction to compensate for shortening. The neutral rotation position is essential. A long trifin nail is inserted over a guide wire under x-ray control. It is started just below the greater trochanter and directed upward through the femoral neck and head and into the pelvis. The hip is filled with small iliac cancellous bone chips. A whole-thickness graft is removed from the dorsum of the ilium and is driven into a slot cut in the bone just above the acetabulum. The other end is laid over a groove cut in the head and the neck and fixed with one screw.

A double plaster spica as far as the toes on the affected side and to just above the knee on the opposite side is applied and retained for a minimum of 4 months. Watson-Jones states that the knee joint upon removal of the plaster is only temporarily stiff but "will become permanently stiff if it is irritated and worried by repeated stretching, passive force, and violence." Regular exercises actively performed throughout the day will regain most of the movement.

[22] Watson-Jones, R., and Robinson, W. C. Arthrodesis of the osteoarthritic hip joint, J. Bone & Joint Surg. 38B:353, 1956.

Femoral Head Prosthesis

The femoral head may be replaced by a prosthesis made of plastic (acrylic, nylon) or metal (stainless steel, Vitallium). Two main types are used. The *cervical* type of Judet, composed of a head piece and a stem, is inserted through the femoral neck and a hole in the lateral cortex.[23, 24] It has been largely discarded in favor of the *intramedullary* type because of frequent sequelae, including loosening, displacement into a varus position, and continued pressure erosion and consequent shortening of the femoral neck. The intramedullary type possesses an elongate stem for insertion into the medullary canal of the shaft.[25] It obtains strong purchase on the bone over a large area, making it less liable to loosen than the Judet prosthesis. For its insertion it is necessary to remove most of the femoral neck. This compromises the possibility of an arthrodesis if the prosthesis should fail. In addition, the prosthesis will not stand up under the requirements of heavy work and often requires a weight-bearing aid. Therefore, the indications are limited.

[23] Judet, R., and Judet, J. Technic and results with the acrylic femoral head prosthesis, J. Bone & Joint Surg. 34B:173, 1952.

[24] MacAusland, W. R.: Femoral head replacement by prosthesis, Surg., Gynec. & Obst. 92:513, 1951.

[25] Thompson, F. R. Intramedullary prosthesis, J. Bone & Joint Surg. 36A:489, 1954.

Indications. The most common indications are a stiff, deformed and painful *osteoarthritic joint, nonunion* of the femoral neck, *aseptic necrosis* of the head and *fresh fractures* of the neck in aged who cannot meet the restrictions imposed by hip nailing. Subcapital fractures are particularly liable to produce aseptic necrosis of the head and therefore are most suitable for prosthetic replacement. This fracture is noted for delay in healing, and the prolonged immobilization is dangerous to the aged patient. A prosthesis quickly restores activity. Conditions which cause *extensive destruction of the head and the neck,* e.g., radium necrosis fractures after radium applications for malignancy of the cervix, should be treated by a prosthesis. When *hip fusion is contraindicated* by lumbar spine disease, prosthetic replacement is an alternative.

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Postoperatively, the extremity is kept in extension and abduction for about 2 or 3 weeks as a precaution against dislocation. Gradually increased and guarded movement in bed is permitted and followed by short periods of graduated weight-bearing. A cane is often

[26] Moore, A. T: The self-locking metal hip prosthesis, J. Bone & Joint Surg. 39A:811, 1957.





FIG. 374. Bilateral congenital subluxation with degenerative changes in the head of the femur and the acetabulum and resulting pain and stiffness. Vitallium mold arthroplasty and Austin Moore prosthesis. (Coventry, M. B.: Proc. Staff Meet. Mayo Clin. 29:48)

necessary. Full unrestricted weight-bearing is permitted after 6 months, or when roentgenograms demonstrate increasing density of bone in and under the prosthesis in stress-bearing locations.

Sequelae.[27, 28, 29] These include, in the order of frequency, dislocation, infection, fracture (of shaft or trochanter), pain, loose prosthesis, broken prosthesis, limp, absorption of neck, erosion of acetabulum, subluxation, progressive loss of motion, calcification of capsule, varus deformity and instability.

These complications are reduced by good exposure, gentle surgical technic, restoration of normal femoral neck length, exact fitting of the prosthesis to the acetabulum, preoperative adductor tenotomies when indicated, capsulectomy, and postoperative maintenance of abduction, particularly by traction.

Principles of Prosthetic Replacement. The following points are essential to attaining satisfactory results:

1. *Metal* withstands wear and tear.
2. *Long intramedullary stem* distributes pressure and provides better fixation
3. *Firm contact with the stump* of the femoral neck over the calcar femorale, which is best suited to receive pressure. Otherwise, progressive resorption of the neck takes place.
4. *Prosthetic head must fit the acetabulum accurately.* Otherwise, the stresses and strains are localized to one point, the acetabulum becomes eroded, and instability is produced.
5. *Adequate length of lever arm* maintains the efficiency of the gluteus medius and reduces the possibility of dislocation.
6. *Anteverted position* of the head.
7. *Postoperative abduction position* for several weeks.
8. *Gradual resumption of motion and weight-bearing.* This will permit reactive bone formation, which will strengthen contact-compression points. Conversely, excessive pressure will effect bone resorption and possibly fracture.
9. *Firm intramedullary implantation.* A loose stem causes intramedullary resorption, further loosening and fracture of the femoral shaft.
10. *Atraumatic surgical exposure.* Division of the abductor muscles produces an abductor lurch and limp and makes postoperative resumption of ambulation very difficult. The posterior approach obviates this complication To avoid formation of osteophytes about the acetabular rim, cutting the capsule at the acetabular attachment is avoided. The use of a bone saw will disseminate bone dust and contribute to calcification of the soft tissues. Sharp dissection and removal of bone by osteotomes reduces the possibility of this complication.

[27] Committee on Scientific Investigation of the American Academy of Orthopaedic Surgeons Preliminary survey on femoral head prostheses, J Bone & Joint Surg. 35A:489, 1953

[28] Mendelsohn, H A, and Alban, S L Complications in replacement arthroplasty of the hip, J Bone & Joint Surg 36A:30, 1954.

[29] Merle d'Aubigne, R., and Postel, M Functional results of hip arthroplasty with acrylic prosthesis, J. Bone & Joint Surg. 36A 451, 1954



FIG. 375. Effect of growth and sex hormone on the epiphyseal plate. (*Left*) *Growth hormone treated:* the plate is thickened mainly by marked proliferation and accumulation of maturing chondrocytes. The layer of hypertrophied chondrocytes is fragile because it is deficient in matrix. (*Center*) *Untreated control:* the layer of maturing chondrocytes is narrow and contributes little to the thickness of the plate. (*Right*) *Treated with sex hormones:* the entire plate is narrow, the chondrocytes lack the orderly columnar arrangement, mature cartilage cells are rare, and bony trabeculae are numerous and thick. Epiphyseal separation is unlikely. (Harris, W. R.: The endocrine basis for slipping of the upper femoral epiphysis, J. Bone & Joint Surg. 32B:5)

SLIPPED UPPER FEMORAL EPIPHYSIS

(Adolescent Coxa Vara; Epiphyseal Coxa Vara; Epiphyseolisthesis)

During the adolescent rapid growth period, the attachments of the upper femoral epiphysis are weakened, and it is displaced downward and backward. The immediate result is a disabling external rotation deformity of the lower extremity. The late effect is degenerative arthritis of the hip.

ETIOLOGY

The actual cause of slipped femoral epiphysis is unknown.

Predisposing factors:

Age. The period of rapid growth from 10 to 17

Sex. Boys are affected predominantly.

Body Type:

1. Fröhlich type of obesity with underdeveloped genitals

2. Long slender rapidly growing individual

Trauma. Usually trivial. The pressure of weight-bearing or muscle contractions superimposed upon the weakened epiphysis is sufficient to cause displacement. Rarely, severe trauma causes epiphyseal fracture dislocation.

Hormone Theory. When the periosteum of a normal epiphyseal plate is removed, the epiphysis can be detached from the shaft by gentle pressure.[30] The line of separation always passes through the layer of maturing hypertrophied cartilage cells in the epiphyseal plate. At this level the chondrocytes are large, and the intercellular matrix, upon which the tissue strength depends, is thin. Anterior pituitary growth hormone will increase the rate of proliferation of chondrocytes and therefore will increase the area of hypertrophied carti-

[30] Haas, S. L: Localization of the growing point in the epiphyseal cartilage plate of bones, Am. J. Orthopedic Surg. 15 563, 1917.

lage cells. This further lessens resistance to pressure. Sex hormone, especially estrogen, inhibits secretion of growth hormone and decreases thickness of the plate and rate of skeletal growth. The hormone also apparently stimulates endochondral bone formation so that newly formed trabeculae are thick and strong. During growth the structure of the epiphyseal plate is dependent upon the relative levels of growth hormone and sex hormone in the circulation. In the adiposogenital syndrome, the genitals are underdeveloped, suggesting that the sex hormone level is low. Consequently, the ratio of growth hormone to sex hormone is large. In the tall thin rapidly growing child, although sex hormone is normal, growth hormone is in excess. Thus is explained the susceptibility of these endocrine types to slipped epiphysis[31] The upper femoral epiphysis is the only one subjected to shearing stresses.

Pathology[32, 33]

The epiphysis slowly displaces inferiorly and posteriorly, the femoral neck shifting upward and rotating anteriorly to the anteverted position. The result is a varus deformity and adduction and external rotation of the femur. Displacement takes place through the layer of hypertrophied mature cartilage cells adjacent to the calcified cartilage layer. The interval produced by separation becomes filled with fibrous tissue, embryonic cartilage and callus, particularly at the posterior and inferior angle. At all times the head remains attached to the neck by soft tissues, particularly the posterior periosteum through which the major vessels pass to reach the epiphysis.

In the early stages, the synovial membrane is swollen, edematous, hyperemic and villous Microscopically, the tissue is hypervascular, and clusters of small round cells surround the vessels. Decalcification and hypervascularity are seen at the junction of the femoral neck and the epiphyseal plate. After several weeks the synovial membrane becomes less vascular and more fibrotic and inelastic. After several months, the epiphyseal junction heals, and the exposed portion of the neck superiorly and anteriorly is covered with fibrocartilage. The head is firmly anchored posteriorly by this new fibrocartilage and thick periosteum. The epiphyseal cartilage closes at the end of the growth period. If displacement persists over years, degeneration supervenes. Aseptic necrosis of the head often results when the posterior epiphyseal attachment and its contained vessels are torn by forceful manipulation and surgical trauma.

Clinical Picture

The onset is very insidious, and the course is slowly progressive. The epiphyseal displacement may vary in degree from slight to extreme. Early symptoms arise when little or no displacement has occurred, although the condition is identified clinically and by certain signs in roentgenograms. This is called the *preslipping stage*. While the epiphysis is separated and gradually shifting backward, the period is designated as the *chronic slipping stage*. When displacement, regardless of degree, has been arrested and the separation has healed, the *stage of fixed deformity* is present. Occasionally, the epiphysis will slip acutely and be completely dislocated. This may be called an *acute traumatic dislocation* of the upper femoral epiphysis. This artificial division into various stages is essential in determining appropriate treatment.

Preslipping Stage. At first, slight discomfort appears about the groin, usually after activity; it subsides with rest. This may be associated with slight stiffness and an occasional limp. Discomfort may radiate along the anterior and medial thigh to the inner aspect of the knee. Symptoms are vague, and no objective findings are apparent.

Chronic Slipping Stage. The *pain* increases in intensity, sometimes acutely, as after a trivial injury such as a misstep. *Limp* is more pronounced and persistent. Objective findings include *tenderness* about the hip and *limitation of motion*, particularly abduction and internal rotation. The limb gradually develops an *adduction and external rotation deformity*.

[31] Harris, W. R. The endocrine basis for slipping of the upper femoral epiphysis, J Bone & Joint Surg. 32B 5, 1950

[32] Howorth, M B · Slipping of the upper femoral epiphysis, J. Bone & Joint Surg 31A 734, 1949

[33] Lacroix, P, and Verbrugge, J Slipping of upper femoral epiphysis; pathological study, J Bone & Joint Surg 33A·371, 1951

Real shortening is due to upward displacement of the femur; *apparent shortening* is due to adduction. As the hip is flexed, the external rotation deformity is accentuated. When slipping is extreme, the gluteus medius is rendered inadequate, and the *Trendelenburg test is positive*. A bilateral severe slipping results in the typical bilateral gluteus medius or *waddling gait.*

Stage of Fixed Deformity. Pain and muscle spasm disappear, but the limp, external rotation and adduction deformity, limitation of internal rotation and abduction, and shortening persist.

Roentgenologic Findings[31]

Anteroposterior and lateral views of both hips are taken. The lateral is taken in the "frogged" position, i e., with the hip flexed to 90° and abducted 45°. The earliest findings in the preslipping stage are a globular swelling of the joint capsule, irregular widening of the epiphyseal line, and decalcification of the epiphyseal border of the metaphysis. As displacement occurs, the head slips inferiorly and posteriorly. The lateral view reveals the slipping more readily than the anterior view. Normally, the epiphysis extends a little above the superior border of the neck. Slight degrees of slipping can be detected by the epiphyseal edge becoming flush with the neck border. The continuity of Shenton's line is broken. Later, the epiphysis slips even further, and the upper medial border of the neck is exposed. This becomes smoothed off. A form of callus fills the interval between head and neck and becomes prominent at the inferior angle. With healing the metaphysis recalcifies, and the line resumes its normal width. Later, it is obliterated completely.

Treatment of Slipped Epiphysis

Conservative Treatment. This consists of freedom from weight-bearing by bed rest, crutches, sling, cast and brace to prevent further displacement. Traction counteracts muscle spasm. Manipulation has been attempted in early cases to reduce displacement. All conservative forms of treatment are in-



Fig. 376. Complete slipped upper femoral epiphysis. Commonly termed acute slipped epiphysis, in reality it is a form of fracture displacement which can also occur at other epiphyseal sites.

effectual. Traction does not reduce the epiphysis, crutches do not prevent pressure on the articular surfaces, and manipulation very often is the cause of a stiff hip. Slipping may continue even at bed rest. Surgery is indicated in all types as soon as the diagnosis is made.[35]

Theoretically, gonadal hormones may be used to limit growth and strengthen the epiphyseal plate. Combinations of estrogen and testosterone may be given parenterally twice weekly during the growth period.

Surgery for Slipped Epiphysis. The aims of surgery are to immobilize, prevent further slip, reduce displacement and effect early closure. Slight displacement requires internal fixation in situ without opening the hip joint by pins, Smith-Petersen nail, or bone pegs. Considerable displacement requires reduction to overcome deformity and limp. A slowly slipping epiphysis, especially when roentgenograms show tissue of some density filling the gap, is firmly fixed and requires considerable manipulative force to reduce. Manipulative procedures are very destructive, the soft tissues including the important posterior peri-

[31] Klein, A., Joplin, R J., Ready, J. A., and Hanelin, J.: Roentgenographic changes in slipped femoral epiphysis, J. Bone & Joint Surg 31A:1, 1943

[35] Cleveland, M, Bosworth, D M, and Hess, W. E. Study displaced capital femoral epiphysis, J. Bone & Joint Surg. 33A 955, 1951.

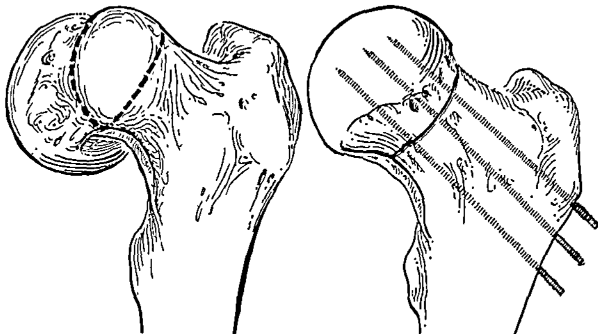


FIG. 377 Correction of old deformity caused by a slipped upper femoral epiphysis. A bony wedge is removed from the juxta-epiphyseal portion of the femoral neck with the base directed anteriorly and superiorly. The epiphyseal plate is drilled, the epiphysis apposed to the neck, and threaded wires are inserted. Note that the posterior periosteum is thick and holds the epiphysis in the displaced position. Forcible traction on the epiphysis will tear this structure and its contained vessels to the epiphysis. (Compere, C. L.: J. Bone & Joint Surg. 32A:351)

osteal attachment are torn, and aseptic necrosis frequently supervenes. A permanently painful and stiff hip results. The severe traumatic displacement in which the epiphysis is completely separated is no exception. Occasionally, reduction of this type may be successful by the Leadbetter maneuver for reduction of a fractured neck, following which blind pinning is done. The maneuver should be performed gently and atraumatically. Otherwise, open reduction and fixation under direct vision are indicated. Opening the capsule anteriorly does not increase the possibility of aseptic necrosis as long as the epiphysis is handled gently.[36] The epiphyseal plate is punctured by multiple drill holes to provide the metaphyseal vessels with rapid access to the epiphysis. When displacement has persisted and become firmly fixed in the deformed position, it is necessary to perform an osteotomy, either through the neck or the trochanteric area. Cervical osteotomy is preferred because accuracy of correction is obtained. Even were it true that this procedure might cause some degree of aseptic necrosis, subtrochanteric osteotomy by contrast is a blind method of correction and results in most cases in a mechanically deficient joint which inevitably must degenerate.



FIG. 378. Slipped upper femoral epiphysis. Anteroposterior view The epiphyseal line is widened and irregular. Callus has filled the interval between the head and the neck and covers the exposed surface of the neck superiorly.

[36] Compere, C L.· Correction of deformity and prevention of aseptic necrosis in late cases of slipped femoral epiphysis, J. Bone & Joint Surg 32A:351-362, 1950.



Fig. 379. Slipped upper femoral epiphysis. Lateral view shows degree of displacement better than anteroposterior view.

Procedure.[37] The hip is exposed through a Smith-Petersen incision. The capsule is incised anteriorly and parallel with the neck. A transverse incision at the acetabular margin widens the capsular opening. If the epiphysis is free and displaced posteriorly, it is elevated with a towel clip. If the epiphysis is fixed in the deformed position, the widened epiphyseal line will present anteriorly. Adjacent to the plate a wedge-shaped section of bone is removed from the neck with its base directed anteriorly and superiorly and the apex inferiorly and posteriorly. The osteotome should not penetrate the intact posterior periosteum. This will enable the epiphysis to be *angulated* anteriorly and superiorly. No attempt is made to pull or shift the head upward. Several drill holes are made in the epiphyseal plate. The epiphysis is apposed to the neck while the leg is rotated internally. Several threaded wires or pins are inserted from below the greater trochanter through the neck and into the head. Improvement of internal rotation and lessening of external rotation should be demonstrated at this point. The capsule is closed; then the wound is closed in layers.

Postoperatively, active motion is started immediately, and crutches are used for several



Fig. 380. Slipped upper femoral epiphysis. Immediate postoperative appearance. (*Top*) Lateral view. (*Bottom*) Anteroposterior view.

[37] Martin, P. H.: Slipped epiphysis in the adolescent hip, J. Bone & Joint Surg 30A 9, 1948.



FIG 381. Slipped upper femoral epiphysis. Two years postoperative. Normal valgus has been restored.



FIG. 382. Slipped upper femoral epiphysis, 2 years postoperative. Lateral view, correction of posterior displacement.

months. The epiphyseal plate is usually obliterated at 4 or 5 months, after which the pins are removed, and weight-bearing is permitted. If one is apprehensive about the possibility of aseptic necrosis, particularly in the completely separated type, weight-bearing is forbidden until the presence of this complication can be determined. Roentgenographic signs of bone necrosis may not appear for many months and are an indication for prolonged protection until replacement by creeping substitution has occurred

TROCHANTERIC OSTEOTOMY. The femur is sectioned similarly to the McMurray osteotomy level. Then the distal fragment is abducted and rotated internally, the amount being determined by the degree of deformity. Blade-plate fixation permits immediate postoperative motion.

COXA PLANA

(Legg-Calvé-Perthes disease;[38, 39, 40] osteochondritis deformans juvenilis; pseudocoxalgia, osteochondrosis of the hips.)

Coxa plana is a self-limited disease of the

[38] Legg, A T An obscure affection of the hip joint. Bost Med & Surg J 162.202, 1910

[39] Calvé, J Sur une forme particulière de pseudocoxalgie greffée sur des déformations caractéristiques de l'extremité supérieure du fémur, Rev de Chir. 30 54, 1910

[40] Perthes, G. C. Uber arthritis deformans juvenilis, Deutsche Ztschr Chir 107.111, 1910



FIG. 383. Legg-Calvé-Perthes disease. Note falsely widened joint space, fragmentation, osteoporosis of metaphysis, shortened and widened femoral neck. (Dr. E. L. Compere's case)

hip in children in which the femoral head undergoes aseptic necrosis and replacement. A variable amount of permanent deformity and restricted motion is the usual result. Together with similar conditions in other epiphyseal regions, it is wrongly classified as an osteochondritis, thereby implying an inflammatory state. Microscopic evidence of inflammation is absent. Osteochondrosis is a more descriptive term.

Etiology

The definite cause of vascular disturbance of the femoral head is unknown.

Predisposing Factors

Age—about 4 to 10.

Sex—males predominantly affected.

Location—either hip, bilateral in 15 per cent.

Trauma and infection are unproved causes. Pituitary and hypothyroid disturbances have been blamed, but these conditions cause retarded, irregular and multiple ossification centers, not aseptic necrosis. Endocrine preparations are of no value in true coxa plana.

Experimental Work

The pathology has been reproduced by ligation of the ligamentum teres and stripping of the periosteum from the femoral neck.[41] Similar but less pronounced changes result from tying the ligamentum teres alone.[42]

Pathology[43, 44]

Obliteration of blood supply produces the picture of aseptic necrosis followed by replacement. The process is best described in stages.

Early Stage. *Bone necrosis*—bony architecture normal, but lacunae are empty. *Marrow degenerate*—amorphous debris fills spaces. *Neighboring bone is hypervascularized* and therefore osteoporotic, particularly in the metaphysis. This constitutes preparation for invasion, removal and replacement of necrotic bone and debris. The diminished density of the osteoporotic bone causes by contrast an apparent increase of density of the necrotic bone. *Overlying cartilage remains viable*—nutrition derived from synovial fluid. Therefore, the gross external appearance of the head is normal. However, the cartilage feels soft and yielding to the touch because of the underlying degenerate tissue.



Fig. 384. Legg-Calvé-Perthes disease. Three years later. Excellent reconstitution.

Regenerative Stage. Necrotic bone is replaced by viable bone. *Capillaries with mononuclear macrophages and foreign body giant cells invade,* absorb debris and fibrose the marrow. *Osteoclasts resorb* necrotic trabeculae. *Osteoblasts form new osteoid. Fragmentation* results from many tonguelike inroads of vessels from the ligamentum teres, the periosteum and the metaphysis via the epiphyseal plate. *Subchondral fractures* of necrotic bone cause multiple minute spicules compressed together.

Healed Stage. Newly formed bone is soft and easily compressed. Flattening of the superomedial aspect, and a cup-shaped or mushroom-shaped contour of the head results. The incongruity of surfaces leads to degenerative arthritis.

In the early stages, the soft tissues about the hip (synovium, capsule, periosteum) are swollen and hyperemic. This constitutes a re-

[41] Miltner, L. J., and Hu. Osteochondritis of the head of the femur; experimental study, Arch. Surg. 27:645, 1933.

[42] Leriche, M. R. Experimental research on the mechanism of formation of osteochondritis of the hip, Lyon chir. 31:5, 1934.

[43] Luck, J. V.: Bone and Joint Diseases, Springfield, Ill., Thomas, 1950.

[44] Haythorn, S. R.: Pathological changes found in material removed at operation in Legg-Calvé-Perthes disease, J. Bone & Joint Surg. 31A:599-611, 1949.

pair reaction and not true inflammation. As this subsides, fibrosis and inelasticity of these tissues restrict motion. The necrotic center by being resorbed at its periphery becomes smaller. In consequence, the area of radiolucency between the bony structure of the center and the acetabulum and between the center and the metaphysis is widened. A false impression is gained of a widened joint space and a thickened epiphyseal plate.

Clinical Picture

The onset is insidious and the course prolonged. About 3 or 4 years elapse before the process has been completed.

Symptoms. *Limp* is the earliest symptom. At first it is slight but gradually becomes pronounced *Pain* is vague and is described as an ache in the groin, the medial thigh and the inner aspect of the knee. It is aggravated by activity and relieved by rest. *Stiffness* may be a complaint.

Findings. *Motion is limited* in all directions, especially abduction and rotation. Pain is experienced at the extremes of motion. Muscle spasm may be apparent in the early stage. A slight degree of muscle atrophy develops. *Tenderness* is elicited over the anterior aspect of the joint.

When the disease has run its course, a moderate amount of restricted motion, slight shortening and an insignificant limp is the usual result. Pain is absent, and the patient is disabled only for activities requiring prolonged standing and running. A picture of degenerative arthritis supervenes years later. Occasionally, reconstitution of the hip and clinical cure is complete.

Roentgenographic Findings[45]

The following findings are characteristic:

1. *Early signs of synovitis*—prominence of soft tissues over capsule
2. *Joint space appears widened*—this is only apparent.
3. *Decreased density of proximal end of metaphysis* due to hypervascularity and osteoporosis
4. *Broadened and shortened femoral neck*
5. *Increased density of head*
6. *Femoral head flattened and widened*
7. *Fragmentation,* then complete decalcification of ossific nucleus as vessels invade and resorb bone
8. *Restored femoral head* varies in size and shape; normal, cup-shaped, mushroom-shaped.

Treatment

The principle involved is *absolute non-weight-bearing during the entire course of the disease.* Complete replacement of the femoral head requires 2 or 3 years. During this time the head is fragile and easily succumbs to pressure. At first, traction is applied to overcome muscle spasm. Bed rest is continued for many months until roentgenograms demonstrate complete regeneration. Ambulatory non-weight-bearing devices are used occasionally. These include crutches, an ischial bearing caliper plus a built-up shoe on the opposite foot and a sling which supports the foot. It is questionable whether these are effective because weight-bearing cannot be completely prevented. Immobilization is contraindicated because it contributes to restricted hip motion The excellence of the final result is proportional to adherence to strict recumbency and the stage at which treatment is started.[46, 47] A poor result consists of pain, limp, disability, shortening of the femur, muscle atrophy, moderate to severe limitation of motion and deformity of the femoral head and neck. A severe degenerative arthritis is inevitable.

Operation[48]

Theoretically, drilling of the epiphyseal plate provides the metaphyseal vessels with easy access to the head, and the course is materially shortened. The results are not convincing.

OSTEOTOMY AT THE HIP

The reasons for osteotomizing the upper end of the femur are:

[45] Ferguson, A. B.: Early roentgenographic changes in Perthes disease *in* Clinical Orthopaedics No. 1, Philadelphia, Lippincott, 1953.

[46] Pedersen, H. E., and McCarroll, H. R.: Treatment in Legg-Perthes disease, J. Bone & Joint Surg 33A 591, 1951.

[47] Pike, M M: Legg-Perthes disease. Method of conservative treatment. J. Bone & Joint Surg 32A. 663, 1950

[48] Howorth, M. B: Coxa plana, J Bone & Joint Surg. 30A:601, 1948.

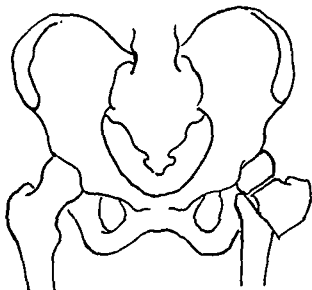


Fig. 385. McMurray osteotomy.

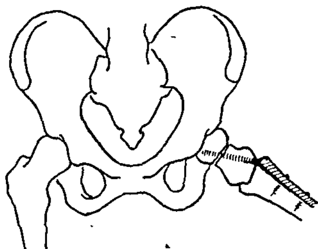


Fig. 386. Dickson geometric osteotomy.

1. To obtain stability. In old unreduced dislocations:

A. *Lorenz bifurcation.* An oblique osteotomy below the lesser trochanter. The upper end of the distal fragment is displaced into the acetabulum. (See Treatment of Congenital Dislocation of Hip)

B. *Shanz.* An osteotomy at the level of the tuber ischii The angle formed by the proximal and the distal portions of the femur should be equal to the angle formed by the completely adducted femur and the mid-line of the body. In a dislocation the upper end of the femur is displaced posteriorly and creates a flexion deformity at the hip The Thomas test measures the degree of flexion which must also be corrected at osteotomy. Therefore, the resulting angle of osteotomy points medially and anteriorly. (See Treatment of Congenital Dislocation of Hip.)

C. *Haas.* An osteotomy done at the level of the acetabulum so that the angle fits into the socket.

2. To obtain a compression force and possible union in ununited fractures of the femoral neck:

A. *McMurray.* The severance is oblique from just below the greater trochanter upward and inward to a point above the lesser trochanter. The distal fragment is displaced medially beneath the femoral head to effect stability. Union between head, neck and distal fragment is accomplished occasionally.

B. *Dickson.* This is a high geometric osteotomy, done for nonunion of the femoral neck. The line of fracture is converted from vertical (shearing force) to horizontal (impacting force). The fracture is fixed by a nail, an osteotomy is done just below the greater trochanter, and the distal fragment is abducted 60° and fixed with a plate. A special osteotome devised by Dickson secures the desired angular section of bone.

3. To relieve pain, particularly in degenerative arthritis. Theoretically, this is accomplished by changing lines of stress. Any of the above procedures are effective. The McMurray type is used most popularly.

4. To correct deformity of coxa vara which follows a slipped upper femoral epiphysis. The epiphysis has displaced to a posterior and inferior position, resulting in restriction of internal rotation motion and shortening of the extremity. A wedge of bone is removed from the neck adjacent to the epiphyseal line, the base of the wedge being anterior and superior.

Pelvic Support Osteotomy for Unreduced Congenital Dislocation of the Hip[49]

This is the operation of choice after the age of 12. In younger patients the osteotomy site tends to straighten, and correction is lost The flexion and adduction deformity should be corrected. Postoperatively, a good range of motion is retained. It is essential that the femur proximal to the osteotomy be movable

[49] Bell, B. T. Pelvic support osteotomy, S Clin. North America 33 1719, 1953.

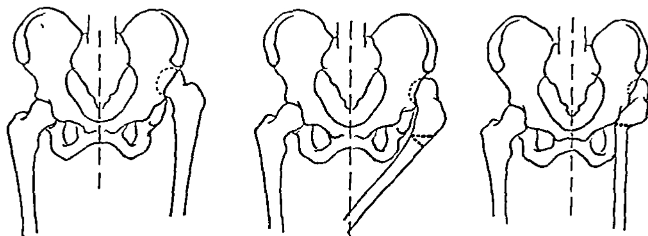


FIG. 387 Pelvic support osteotomy for irreducible congenital dislocation of hip. (*Left*) Unreduced congenital dislocation. (*Center*) Extremity completely adducted to determine angle of abduction after osteotomy. (*Right*) Final position after osteotomy.

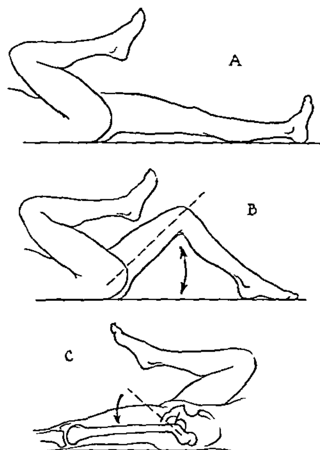


FIG. 388 Osteotomy for correction of flexion deformity of hip. (A) Hip remains fully extended normally when opposite hip is fully flexed. (B) Flexion of opposite hip will reveal flexion deformity. The degree is measured by the angle formed between the affected thigh and the surface of the examining table. (C) Final correction by osteotomy.

and not fixed. If the head is fixed, e.g., after a previous operation or in osteoarthritis, the head and the neck of the femur are removed, and the pelvic support osteotomy is done at a later date.

First, the angles are determined. An anteroposterior roentgenographic view of the pelvis is taken while the limb is held in marked adduction. The angle which the femur forms with the mid-line of the body is measured Then the degree of flexion deformity of the hip is ascertained. Both angles must be corrected at operation. The osteotomy site is selected so that the angle will protrude just under the acetabulum.

PIGMENTED VILLONODULAR SYNOVITIS (XANTHOMATOSIS) OF THE HIP JOINT[50, 51]

This term is used to describe all xanthomatous tumors of joints, and refers to their gross appearance. True synovial tumors are rare, and the most commonly reported are the xanthomatous tumors. The most common site is in the knee, but other joints have been involved.

[50] Ghormley, R K., and Romness, J. O.: Pigmented villonodular synovitis of the hip joint, Proc. Staff Meet. Mayo Clinic 29:171, 1954

[51] Jaffe, H L., Lichtenstein, L., and Sutro, C. J.: Pigmented villonodular synovitis, bursitis, and tenosynovitis; discussion of synovial and bursal equivalents of tenosynovial lesions commonly denoted as xanthoma, xanthogranuloma, giant cell tumor or myeloplaxoma of tendon sheath, with some consideration of this tendon sheath lesion itself, Arch Path. 31:731. 1941.

Clinically, villonodular synovitis of the hip is characterized by progressively increasing dull, aching pain and limitation of motion. Findings of a limp and a positive Trendelenburg sign point toward the hip where a flexion contracture may be found. Attempts to exceed the range of motion passively are very painful.

X-ray study reveals monoarticular involvement, narrowing of the joint space, irregular joint surface changes, and cystic areas in the acetabulum, the head and the neck of the femur. The findings in degenerative arthritis, on the other hand, are usually limited to the head of the femur.

Laboratory findings are not diagnostic. Hypercholesteremia may or may not occur.

Grossly, at operation, the tissue looks shaggy and thick, and strandlike bits of soft brownish-red tissue with intermittently scattered yellowish gold coloring are seen.

Microscopically, the villi are lined by 1 to 4 layers of synovial cells which frequently contain pigment. Thin-walled vascular channels course through the stroma which is loose. There are scattered collections of pigment-filled stromal cells, multinucleated giant cells of foreign-body type and hemosiderin and lipoid-bearing foam cells. A variable amount of hyalinized connective tissue is present. The consensus at the present time is that the lesion is benign and probably of inflammatory nature.

Treatment is by total excision followed by x-ray therapy.

TRANSIENT SYNOVITIS OF THE HIP[52, 53, 54]

(Transitory Coxitis; Coxitis Cerosa Seu Simplex; Acute Transitory Epiphysitis)

This consists of a very commonly seen temporary nonspecific inflammation of the synovium of the hip in children usually occurring without apparent cause. Less often it follows trauma or is associated with allergy, tonsillitis, or exanthems.

The onset is insidious. The child limps and complains of pain about the hip, the thigh, or the knee. The extremity is held in the protective attitude of flexion, adduction and internal rotation. Passive motion is restricted by muscle spasm. Tenderness is elicited over the hip. The temperature is normal or low grade, rarely high.

X-ray examination at first glance is normal. However, the soft tissues overlying the intrapelvic aspect of the acetabulum are swollen and form a prominent shadow. This is known as the *obturator sign.* Other laboratory tests are negative.

The condition is readily differentiated from other disease, such as tuberculosis, rheumatic fever, etc., by the rapidity with which it subsides with a few days of bed rest.

Treatment consists of bed rest and traction to overcome the painful muscle spasm.

Occasionally, changes in the metaphysis adjacent to the epiphyseal plate are seen in roentgenograms. This supposedly constitutes an infective embolus from a distant focus, particularly the tonsils, with which it is often associated. The duration is a few weeks to a few months, and cure is often dramatically effected by tonsillectomy.

CALCIFIED TENDINITIS OF THE HIP JOINT[55, 56, 57]

Amorphous calcium deposits in the tendons about the hip are exactly comparable with calcified tendinitis about the shoulder. The soft toothpastelike or chalky material forms in the gluteus medius tendon lateral to the greater trochanter and in the gluteus minimus tendon superior to the capsule.

Clinically, severe pain appears acutely, the patient limps, and the limb is held protectively in flexion, abduction and external rotation. Muscle spasm limits motion in all directions. Tenderness is detected over the site of the deposit.

[52] Edwards, E. G : Transient synovitis of the hip joint in children: report of 13 cases, J.A.M.A 148:30, 1952

[53] Miller, O. L.: Acute transient epiphysitis of the hip joint, J.A.M.A. 96:575, 1931.

[54] Lucas, L.: Painful hips in children, Am. Acad. Orthop. Surgeons, Lect., vol 5, 1948.

[55] Goldenberg, R. R., and Leventhal, G. S.: Supratrochanteric calcification, J. Bone & Joint Surg. 18: 205, 1936.

[56] Jones, G. B.: Acute episodes with calcification around the hip joint, J. Bone & Joint Surg. 37B:448, 1955.

[57] Spear, I M., and Lipscomb, P. R : Noninfectious trochanteric bursitis and peritendinitis, S. Clin. North America 32:1217, 1952.

X-ray films reveal the cloudy opacity in the soft tissues.

Treatment

Conservative measures such as rest, heat and x-ray therapy are effective. Infrequently, surgical excision is necessary. Often the deposit may be broken up with a needle under local anesthesia, following which it is absorbed.

INTRAPELVIC PROTRUSION OF THE ACETABULUM[58-61]

(Protrusio Acetabuli; Arthrokatadysis; Otto Pelvis)

Abnormal deepening of the acetabulum when not caused by trauma is relatively rare. The mechanically deficient joint gradually undergoes degenerative changes, resulting in a rigid disabling hip flexion deformity.

Etiology

Idiopathic Type: This develops during childhood or adolescence and remains unrecognized a long time until degenerative arthritis supervenes. The condition is bilateral and occurs mainly in women.

Type Due to Antecedent Disease or Trauma

Clinical Picture

The onset varies considerably. Usually, at about middle age the complaint of *stiffness* arises. *Pain and limp* appear as osteoarthritis develops. *All motions* become progressively more *limited*, especially abduction and external rotation. A *hip flexion deformity* secondarily causes increased lumbar lordosis, pelvic rotation and shortening. *Rectal examination* reveals a globular mass on the lateral rectal wall. The pain is increased by weight-bearing and becomes progressively worse.

Roentgenologic Findings

The acetabular wall protrudes medially. The normal pear or teardrop figure is altered. The teardrop is composed of the intersection of 3 lines. The lateral line is continuous with the acetabular floor. The medial line is the inner pelvic wall. The bottom line is formed by the anterior obturator tubercle. When the medial and lateral lines reverse their positions, an abnormally deep socket is present. The CE angle of Wiberg may be used to detect the early case. Protrusio acetabuli exists when the angle approaches or is larger than 50° (normal average 36°). At first the head is not deeply placed, because the socket is filled with fat and areolar tissue. Later, as the femoral head migrates inwardly degenerative changes appear.

Treatment

Early symptoms are changed by relief from weight-bearing and traction. When degenerative disease and deformity are disabling, surgical intervention is necessary. When an adequate thickness of acetabular wall is present, arthrodesis is indicated. If the condition is bilateral, preservation of motion is paramount It is necessary to strengthen the acetabulum first by placing a semilunate portion of the femoral head in the deepest part of the socket. This will fuse to the acetabular wall. This is followed by insertion of a prosthesis.[62]

BURSITIS ABOUT THE HIP

As many as 18 bursae about the hip have been described. Only 3 are of importance: the trochanteric, the iliopectineal and the ischiogluteal. Being physiologically and developmentally related to tendon sheaths and synovial membranes, the bursae are subject to similar diseases, namely, traumatic inflammation, infection, benign growth (pigmented villonodular synovitis), malignant growth (synovioma) and gout. The usual bursitis is an inflammation which is reactive to overuse or excessive pressure and subsides with rest, hot fomentations and needle puncture Corticosteroids and ACTH are rarely necessary.

Trochanteric Bursa

This large bursa lies between the tendon of

[58] Otto, A W In Pfannenbeckenmisstaltung Infolge deformierender Osteoarthritis neue seltene Beobachtungen zur Anatomie Physiologie und Pathologie gehorig Berlin, Rucker, 1824

[59] Ghormley, R K, and Dockerty, M B Arthrokatadysis (Otto pelvis), Surgery 29 255, 1951

[60] Friedenberg, Z B Protrusio acetabuli, Am J. Surg. 85:764, 1953

[61] Gilmour, J · Adolescent deformity of the acetabulum An investigation into the nature of protrusio acetabuli. Brit. J. Surg. 26 670, 1939

[62] Lipscomb, P. R : Surgical repair of acetabular defects, Proc Staff Meet Mayo Clin. 29 53, 1954



FIG. 389. Arthrokatadysis (Otto pelvis). The intrapelvic protrusion of the acetabulum is congenital and bilateral.

insertion of the gluteus maximus and the posterolateral prominence of the greater trochanter. Tenderness is elicited by pressure or by tensing the muscle when the hip is flexed and rotated internally. Rarely, a tuberculous infection can involve this bursa.

Treatment. Purulent infection demands incision and drainage. The incision is made just behind the trochanter with due regard for the adjacent sciatic nerve. Tuberculosis requires excision of the bursa plus streptomycin and chemotherapeutic agents.

ILIOPECTINEAL BURSA

This is the largest and most constant bursa about the hip. It lies between the iliopsoas muscle anteriorly and the iliopectineal eminence posteriorly. It is situated lateral to the femoral vessels and overlies the capsule of the hip When inflamed, the pain and the tenderness are in the lateral part of Scarpa's triangle; sometimes the swelling is sufficient to obliterate the inguinal groove. Irritation of the adjacent femoral nerve causes pain referred along the anterior thigh as far as the upper inner aspect of the leg. Tensing the iliopsoas by contraction, as when actively flexing the hip, or by stretching the muscle, as when extending the hip, will increase the pain. Very frequently, the bursa communicates with the joint cavity and is involved simultaneously with the synovium. Pigmented villonodular synovitis can involve the iliopectineal bursa and enlarge and distend the latter so that a

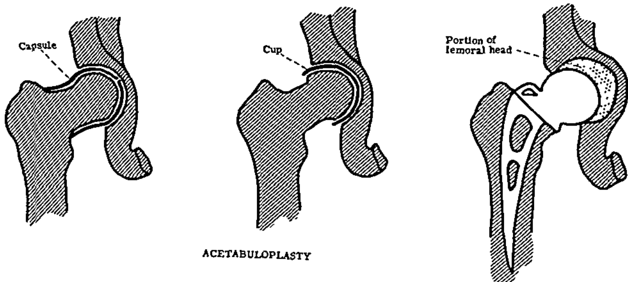


FIG. 390. Acetabuloplasty for protrusio acetabuli. Cup arthroplasty is preferred when sufficient thickness of the acetabular wall is present. (Lipscomb, P. R.: Proc. Staff Meet. Mayo Clin. 29:53)

large tumorlike process may extend upward intra-abdominally behind the psoas muscle.[63, 64]

Treatment. Proper exposure of the bursa is essential for excision or for drainage for infection. The incision begins at the anterior superior iliac spine and extends along the *medial border of the sartorius for several* inches. The superficial circumflex iliac vessels are ligated and divided. The sartorius is severed near its tendinous origin and retracted laterally, the femoral nerve is retracted medially The rectus femoris is severed near the *anterior inferior spine and retracted. The iliopsoas is now exposed. The bursa beneath* the muscle is exposed when the muscle is relaxed by flexing and rotating the thigh externally; or more complete exposure is obtained by severing the tendon of insertion near the lesser trochanter. Because of the frequency of communication with the joint, infections demand simultaneous drainage of the hip joint. When growths are present, the interior of the joint should be explored

Ischiogluteal Bursa

This bursa overlies the ischial tuberosity and becomes inflamed in occupations demanding prolonged sitting (tailor's or weaver's bottom). Irritation of the adjacent sciatic nerve produces symptoms of sciatica.

Treatment. Avoidance of pressure generally relieves symptoms. For incision or removal, a transverse incision is made along the lower border of the gluteus maximus over the tuberosity. The sciatic nerve runs just lateral to the tuberosity and should be exposed and protected.

SNAPPING HIP

An audible, palpable, or visible snap is made by a taut fascial band which suddenly slips over the prominence of the greater tuberosity when the hip is flexed, adducted, or rotated internally. The condition usually is painless but may become painful when an underlying bursa becomes inflamed. The band consists of the thickened *posterior border of* the iliotibial band or anterior border of the tendinous insertion of the gluteus maximus muscle. Only when this condition constitutes a source of annoyance should treatment be undertaken. Under local anesthesia the offending fascial band can be demonstrated as the *patient performs the particular movement* necessary to produce the snap. The band is severed and left unsutured; or the cut fascia may be sutured anteriorly or posteriorly on itself or sutured directly to the greater trochanter. Immediate exercise postoperatively is done.

Other conditions may cause clicking about the hip and must be differentiated from snapping hip. These include osteochondromatosis, a jagged edge on the articular surface caused by fracture or degenerative arthritis, and subluxation, either congenital or associated with weakened hip musculature.

TUBERCULOSIS OF THE HIP

Pathology

When the infection starts in the epiphysis or the neck of the femur, the exudative process causes diffuse decalcification of the upper end of the femur. Because the head and the *neck of the femur are entirely intracapsular,* [illegible] increased intra- [illegible]

tion and external rotation. As the capsule thickens by fibrosis and contracts, the position becomes one of flexion, adduction and internal *rotation. The destruction spreads to involve* the acetabulum. Destruction may be sufficiently great to cause a pathologic dislocation Because the acetabular floor is thin, it is perforated fr[illegible] of an intra-pelvic abs[illegible] and the [illegible] *hip or down the thigh.*

When infection starts in the pelvis about the acetabulum, the joint involvement and therefore the symptoms are late. In consequence, by the time the case is first seen, the bone is beyond the stage of exudation and in a state of caseous destruction.

Healing without surgical intervention is by fibrous ankylosis The fibrous walled cavities enclose lesions which at any time can become reactivated and cause further destruction and

[63] Waisser, J R, and Robinson, D W Pigmented villonodular synovitis of the iliopectineal bursa, J. Bone & Joint Surg 33A 988, 1951

[64] Carr, C. R., Berly, F V, and Davis, W. C. Pigmented villonodular synovitis of the hip joint: a case report, J. Bone & Joint Surg 36A 1007, 1954.

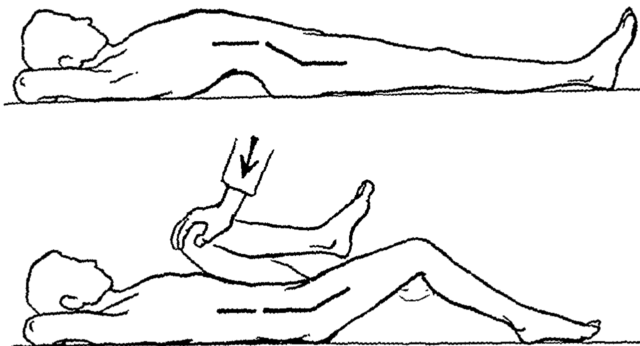


Fig. 391. The Thomas test for flexion contracture of the hip. With the lower extremities fully extended, the pelvis is rotated forward, causing lumbar lordosis. By flexing the uninvolved hip, the pelvis is restored to the horizontal position, the contracted hip pulls the thigh forward, and the lumbar curve is obliterated. The angle which this thigh forms with the surface of the table designates the degree of flexion contracture.

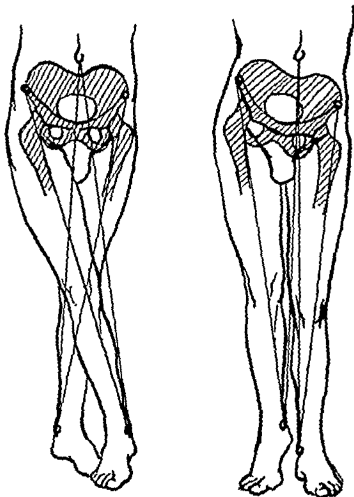


Fig. 392 Adduction deformity at the hip When the legs are kept parallel as when walking, the pelvis displaces upward on the adducted side and causes apparent shortening However, measurement of limb length from the anterior superior iliac spine to the medial malleolus of the ankle will reveal no loss of length.



Fig. 393 Tuberculosis of the hip. Treatment by arthrodesis.

dissemination. Final and complete healing is by bony ankylosis. In spite of firm osseous fusion, with further growth there is a marked tendency to adduction deformity

Tuberculosis of the hip frequently causes premature fusion of the lower femoral epiphyseal plate with consequent shortening of several inches. The prolonged immobilization necessitated in treatment also contributes to this condition. (For further discussion on etiology and pathology, see Tuberculosis in section on Infections.)

Clinical Picture

The onset is usually insidious, although occasionally it may be acute. Children are affected most commonly.

Symptoms and Findings

Pain about the hip occurs, particularly *with weight-bearing* and often is *referred to the inner aspect of the knee* (where it may be the only presenting symptom). *Motion is limited in all directions.* Early, the extremity is held in *flexion and abduction,* causing apparent lengthening. Later, *flexion and adduction* develop, causing apparent shortening.

The *Thomas test* reveals a *flexion contracture* of the hip. This is performed by flexing the unaffected hip enough to obliterate the lumbar lordosis and fix the pelvis. Normally, it is possible to extend the opposite hip completely so that the thigh touches the table. When the hip is contracted in flexion, its severity is measured by the angle which the incompletely extended thigh forms with the table.

In the standing position the flexed hip rotates the pelvis forward and accentuates the lumbar *lordosis.* Abduction and adduction elevate and depress the pelvis, respectively, and thereby cause *scoliosis* of the lumbar spine.

During the acute stage *muscle spasm* is severe. At night the spasm relaxes, permits movement, and resultant pain causes the typical *night cries.* The child resists attempts to move the hip. As the acute stage subsides, pain and spasm are less, and severe generalized *muscle atrophy* of the entire lower extremity develops. Actual *shortening* becomes obvious with passage of time. *Abscesses and sinuses* may present.

Treatment

Proponents of conservative treatment use traction to overcome deformity followed by prolonged immobilization. This at best results in fibrous ankylosis. Lesions although healed by scar are subject to reactivation at any time Destruction and deformity may continue in spite of immobilization and traction. Continued prolonged immobilization may cause extreme atrophy and greatly reduced growth Conservative treatment should be used only for constitutional improvement preliminary to surgery. During this time traction will lessen pain.

The surgical aim is osseous fusion. Intra-articular fusion is difficult to obtain because of poor vascularity of tuberculous tissue and an excess amount of cartilage in children However, resection of the abnormal tissue will expose good cancellous bone and permit correction of deformity. This is supplemented by [illegible] which a bony [illegible] to the ilium [illegible] Previous to [illegible] surgery was contraindicated for fear of introducing sec-

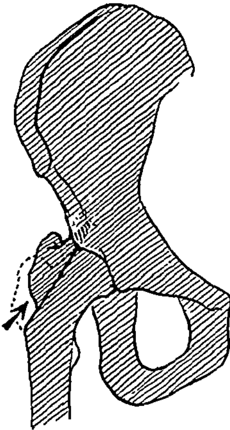


FIG. 394. The Hibbs arthrodesis.

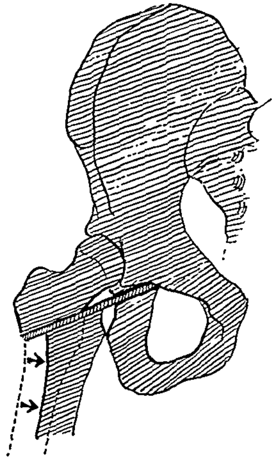


FIG. 395. The Brittain ischiofemoral arthrodesis. The femoral shaft is displaced medially, and the graft is inserted into the ischium.

ondary infection. Now surgery is done even in the presence of abscesses and draining sinuses.

After fusion, with further growth, an adduction deformity practically always develops. This can be prevented by intrapelvic obturator neurectomy. After it develops, it can be corrected by subtrochanteric osteotomy.

Intra-articular Arthrodesis. Resection of cartilage and granulation tissue down to normal-appearing bone creates an interval with poor surface contact. The success of arthrodesis demands firm contact compression. Therefore, multiple autogenous cancellous bone chips are packed into the joint. The femur is placed in sufficient abduction to compensate for shortening, and even further to compensate for expected adduction. In abduction the adductor muscles are taut and aid in approximating joint surfaces. The hip is flexed slightly, and the limb is in neutral rotation. A plaster cast is applied from the nipples to the toes on the affected side and to just above the knee on the opposite side. After 2 or 3 months union may be sufficient to allow some weight-bearing, which helps to stimulate further union. Immobilization is maintained for at least 15 months.

Intra-articular arthrodesis produces a much lower percentage of successful fusions than by the extra-articular or combined procedures.[65]

Extra-articular Arthrodesis. These are of 2 types: the iliofemoral and the ischiofemoral. Hibbs described an iliofemoral fusion by which the greater trochanter is removed and used to bridge the iliofemoral interval.[66] Other procedures utilize bone grafts from the ilium or the tibia. The effectiveness of this procedure is reduced by a distraction force imposed on the graft by the strong adductor pull on the hip. Trumble introduced the ischiofemoral fusion in which a strong tibial cortical graft

[65] Ahlberg, A: Radical operative treatment of the tuberculous hip, J Bone & Joint Surg 30A:550, 1948.

[66] Hibbs, R A: A preliminary report of 20 cases of hip joint tuberculosis treated by operation fixing the joint, J. Bone & Joint Surg. 8:522, 1926.



Fig 396. Tuberculosis of the hip.



Fig 397. Tuberculosis of the hip. Postoperative appearance. Hip fused in abduction to compensate for shortening.



Fig. 398. Tuberculosis of the hip 2½ years postoperative.



FIG. 399. Tuberculosis of the hip 4 years postoperative. Trabeculae are continuous from pelvis to femur. Position of abduction maintained.

is wedged between the ischium and the trochanteric area.[67] This takes advantage of the adductors
Brittain
chanteric
fragment medially against the ischium.[68] This has greatly increased the percentage of successful fusions

Hibbs Iliofemoral Procedure. A longitudinal incision starts at a point on the iliac crest 2 inches posterior to the anterior superior iliac spine and extends distally over and beyond the greater trochanter. Exposure is between the fibers of the tensor fascia femoris and, at a deeper level, of the gluteus medius and minimus, exposing the superior aspect of the joint capsule. The base of the trochanter is exposed subperiosteally. The anterior three fourths of the trochanter is detached with a chisel, leaving undisturbed the muscle attachments at the superior pole. The capsule is split and partially excised, revealing the superior aspect of the femoral neck from which the cortex is removed. The supra-acetabular portion of the ilium is elevated with a chisel. The trochanter is rotated so that the distal end is transposed beneath the elevated iliac flap, its undersurface fitted closely to the cancellous bone of the femoral neck. Suturing of the trochanter to the soft tissues and abducting the thigh holds the graft snugly against the ilium.

Brittain Ischiofemoral Procedure. The lateral aspect of the femur is exposed. Just below the great trochanter, a drill bit is inserted parallel with the floor and at an angle of 45° with the shaft. It penetrates the femur and

[67] Trumble, H. C.: Fixation of hip joint by an extra-articular bone graft, Australian & New Zealand J. Surg. 1:413, 1932.

[68] Brittain, H. A.: Ischiofemoral arthrodesis, Brit. J. Surg. 29:93, 1941.



FIG. 400. Tuberculosis of the hip.

enters the ischium. The position is verified by roentgenograms. A large tibial graft is beveled at one end. An oblique osteotomy is done by inserting an osteotome along the guide pin until it enters both cortices of the ischium. If the posterior edge of the chisel is in front of the posterior margin of the great trochanter and if the chisel is held horizontally, the sciatic nerve is avoided. The osteotome is levered up and down to widen the ischial opening. Next, the graft is inserted along the osteotome and driven into the ischium. The distal fragment is displaced medially by direct pressure and by abduction. Finally, the osteotome is removed. After closure a one-and-a-half spica cast is applied.

25

The Knee

SURGICAL ANATOMY

The knee joint is the largest articulation in the body. For stability, it depends upon the capsule, the collateral and the cruciate ligaments and surrounding muscles.[1]

Capsule

A fibrous structure completely invests the joint particularly on its posterior aspect, which is strengthened by the oblique popliteal ligament. Laterally, it is reinforced by a thickening which bridges the penetrating tendon of the popliteus muscle. Additional support is provided by the collateral and the cruciate ligaments. The anterior capsule is composed of the quadriceps tendon, the patella, the infrapatellar tendon, and the blending of the fibrous aponeuroses of the vasti muscles.

Synovia and Bursae

The synovial membrane lines the inner aspect of the capsule. It extends upward on the anterior aspect of the joint under cover of the quadriceps tendon to form the suprapatellar pouch. A diverticulum is prolonged posteriorly and distally on the popliteus tendon. Another pouch extends posteriorly to communicate with a bursa lying between the semimembranosus and the inner head of the gastrocnemius. The cruciate ligaments are enveloped in synovium, which renders them extrasynovial. The infrapatellar fat pad, triangular in shape, lies between the distal part of the patella and the upper, anterior margin of the tibia. It is covered by a fold of synovial membrane, the lateral free margins of which extend to the intercondylar notch and are known as the ligamenta alaria. The central portion of the fold is known as the ligamentum mucosum.

[1] Abbott, L. C., and Carpenter, W. F.: Surgical approaches to the knee joint, J. Bone & Joint Surg 27:3, 1945.

The semilunar cartilages (menisci) lie between the outermost margins of the articulating surfaces of the femur and the tibia. They are wedge-shaped structures, free at their inner margins but attached to the tibia at their outer margins by the coronary ligaments. Each cartilage presents two extremities or horns, which are fixed by fibrous attachments to the intercondyloid eminence on the upper surface of the tibia.

Numerous bursae have been described. The following are the most constant and of surgical importance. On the anterior aspect of the knee, the prepatellar bursa lies between the skin and the patella. The superficial infrapatellar bursa is between the skin and the patellar tendon, while the deep infrapatellar bursa lies deep to the tendon. On the lateral side of the joint, there are usually 4 bursae. One lies between the lateral head of the gastrocnemius and the capsule; another between the fibular collateral ligament and the biceps tendon; a third between the fibular collateral ligament and the popliteus tendon; and the fourth, usually a synovial extension from the joint, lies between the popliteus tendon and the lateral femoral condyle. On the medial side of the joint, a bursa, often communicating with the joint, lies between the medial head of the gastrocnemius and the tendon of the semimembranosus. A second bursa lies superficially between the tibial collateral ligament and the tendons of the sartorius, the gracilis and the semitendinosus. A third is located beneath the tendon of the semimembranosus at its attachment to the head of the tibia. Additional bursae are located beneath the tibial collateral in relation to the meniscus and the capsule.

Blood Supply

The blood supply is furnished by the genicular branches of the popliteal artery, the arteria genu suprema, a branch of the fem-

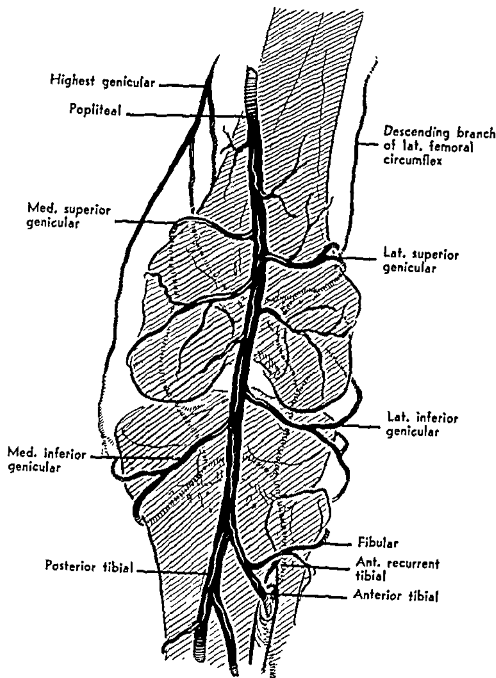
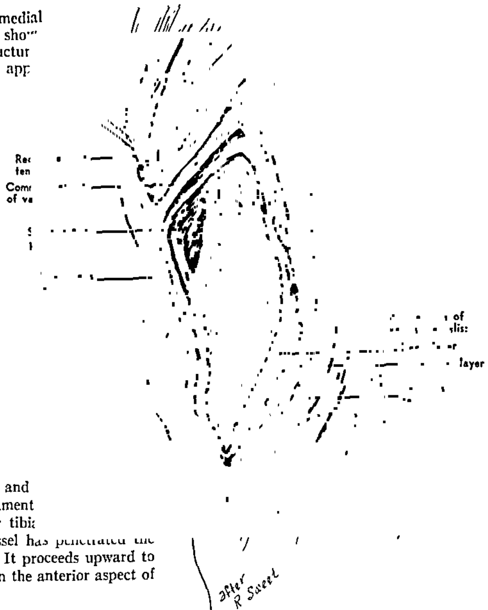


FIG. 401. Arterial anastomosis about the knee. Note that the lateral inferior genicular artery directly overlies the joint margin where it may be injured in approaches to the lateral meniscus. The extensive collateral network makes it possible to obliterate the popliteal without jeopardizing the distal portion of the extremity.

oral artery, the descending branch of the lateral femoral circumflex artery, and the recurrent branch of the anterior tibial artery. The terminals of these vessels form a rich network and are arranged in 3 arterial arches over the anterior aspect of the joint. The uppermost of these arches lies in the midst of the superficial fibers of the quadriceps muscle, near the upper border of the patella. The 2 lower arches are directed transversely through the fatty tissues behind the patellar tendon. The superior medial and lateral geniculate arteries are given off by the popliteal artery proximal to the intercondylar notch. They pass inward and outward beneath the hamstring tendons directly upon the femur, pierce the inner and the outer intermuscular septa and anastomose in the medial and the lateral vasti with the arteria genu suprema and the descending branch of the lateral femoral circumflex. The inferior medial and lateral geniculate arteries are given off from the popliteal artery at the level of the lower border of the oblique popliteal ligament. They pass inward and outward close to the bone. The inferior medial artery passes along the upper border of the popliteus muscle and beneath the tibial collateral ligament. The inferior lateral geniculate vessel passes along the upper border of the fibular head of origin of the soleus, around

Fig. 402. The anteromedial aspect of the knee joint, sho- ing the component structur making up the extensor app ratus.



the neck of the fibula and fibular collateral ligament branch of the anterior tibi off after the main vessel has penetrated the interosseus membrane. It proceeds upward to join the anastomosis on the anterior aspect of the joint.

Cutaneous Nerves

These nerves are important because their severance may cause permanent impairment of sensation. Painful neuromata may form. The infrapatellar branch of the saphenous nerve is divided most frequently, causing anesthesia about the tibial tubercle. The nerves form anatomic landmarks and guides to deeper structures. For example, the posterior cutaneous nerve of the calf can be traced to its origin as the first branch given off from the posterior tibial nerve. The nerves supplying sensation to the knee are:

1. Anterior division of the lateral cutaneous nerve of the thigh
2. Intermediate cutaneous nerve of the thigh
3. Anterior and posterior divisions of medial cutaneous nerve of thigh
4. Saphenous nerve
5. Posterior cutaneous nerve of thigh
6. Posterior cutaneous nerve of calf
7. Lateral cutaneous nerve of calf (from peroneal)
8. Anastomotic peroneal nerve of leg (from peroneal)

Anterior Aspect of the Knee Joint

The quadriceps tendon is subdivided into 3 parts. The anterior portion is the tendon of the rectus femoris, the intermediate portion is the common tendon of the vastus lateralis and the vastus medialis, and the posterior portion is the tendon of the vastus intermedius The genu articularis tendon joins the tendon of the vastus intermedius and provides a slip which inserts into the apex of the suprapatel-

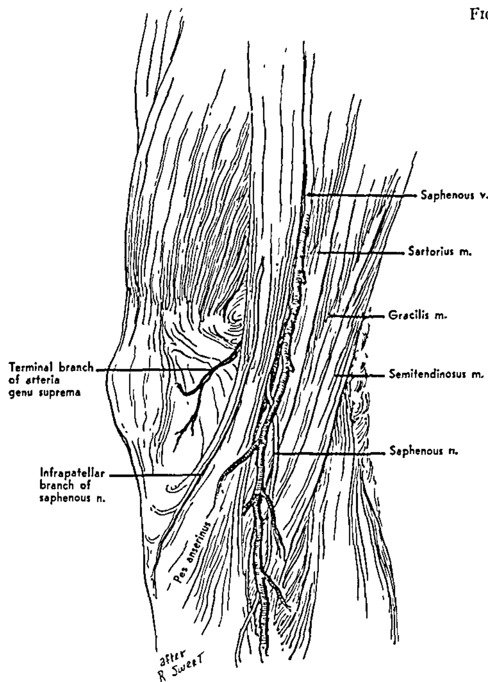


Fig. 403. Medial aspect of the knee.

lar synovial pouch. Between the adjacent margins of the vasti, a fascial sling is formed, which acts as a cradle for the rectus tendon.

The aponeurosis of the vastus medialis distally divides into 2 distinct layers over the anteromedial aspect of the knee. The anterior layer passes across the patella and blends with the aponeurosis of the vastus lateralis. Below the level of the tibial tubercle it becomes continuous with the deep fascia of the leg. The posterior or deep layer of the aponeurosis of the vastus medialis is inserted into the medial border of the patella and the upper margin of the tibia. The synovial membrane lies behind this layer. On the outer aspect of the knee, the deep fascia and the aponeurosis blend to form a single layer which is inserted into the lateral margin of the patella and the upper border of the tibia. This layer fuses with the iliotibial band and the fascia enclosing the biceps tendon.

On the anteromedial surface, 4 layers of fascia are found, namely, the thin superficial fascia, the fascia lata, sometimes blended with the anterior layer of the aponeurosis of the vastus medialis, and the deep layer of the aponeurosis of the vastus medialis. On the anterolateral aspect of the joint, the blending of the

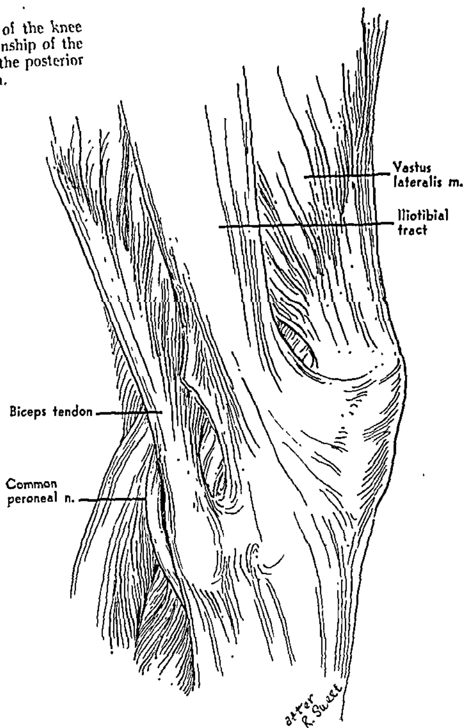


Fig. 404. Lateral aspect of the knee joint. Note the close relationship of the common peroneal nerve to the posterior border of the biceps tendon.

fibrous aponeurosis with the iliotibial tract is so intimate that only 2 layers are distinguishable—the thin superficial fascia and a deep layer of deep fascia and fibrous aponeurosis

With the knee in extension, the quadriceps tendon forms an obtuse angle with the patella and the patellar tendon of approximately 170°. Therefore, curvilinear incisions are preferable. With the knee in flexion, the iliotibial tract and the biceps tendon stand out prominently as they pass to their respective insertions into the lateral margin of the tibia and the head of the fibula. On the posteromedial surface of the joint, the tendons of the sartorius, the gracilis and the semitendinosus are seen where they insert into the medial aspect of the upper end of the tibia.

Medial Aspect of the Knee Joint

The tibial collateral ligament is an extensive structure which may be divided into 4 descriptive portions. The anterior superficial portion is a flat band, approximately 4 inches in length and ½ inch in width, attached above

to the medial epicondyle of the femur and below, about 1½ inches distal to the joint line, to the medial surface of the tibia. The deep anterior portion lies beneath and is intimately blended with the superficial part. It consists of short fibers which extend from the medial epicondyle to the margin of the tibial condyle. The posterior superior oblique portion extends from the posterior part of the femoral attachment downward and backward to the tibial margin, where it gains an extensive attachment This portion helps to anchor the medial meniscus. The posterosuperior and the posteroinferior oblique portions of the ligament blend with the posteromedial portion of the joint capsule, forming a hemispherical pouch. This pouch is lax on flexion of the knee, but in extension it becomes tense as it encloses the posterior part of the medial femoral condyle. This aids stabilization of the knee in extension.

The medial intermuscular septum extends from the fascia lata inward and attaches to the linea aspera. It blends with the tendon of the adductor magnus, which inserts on the adductor tubercle of the femur. This membrane separates the vastus medialis in front from the adductor magnus and the adductor longus behind. It separates the popliteal space from the anterior compartment of the knee. The tendons of the sartorius, the gracilis and the semitendinosus embrace the posteromedial aspect of the medial femoral condyle, and, as they pass to their insertions in the proximal part of the anteromedial surface of the upper part of the tibia, they are separated from each other by a bursa. The tendons span the inferior portion of the tibial collateral ligament The internal saphenous vein passes along the posteromedial aspect, lying on the tendon and the muscle belly of the sartorius

Two branches of the medial cutaneous nerve of the thigh are found. The anterior branch crosses the sartorius muscle and emerges from the fascia lata in the distal part of the thigh in front of the internal saphenous vein. The posterior branch runs along the posterior border of the sartorius, emerges from the fascia lata on the medial side of the knee and lies behind the sartorius tendon and the saphenous nerve. The saphenous nerve is the longest branch of the femoral nerve. It emerges from the lower end of Hunter's canal by passing beneath the fibrous expansion which stretches between the vastus medialis and the adductor muscles. It is accompanied by the saphenous branch of the arteria genu suprema. As it emerges from under cover of the sartorius muscle, it penetrates the deep fascia and gives off the infrapatellar branch, which pierces the sartorius and appears on the surface of the fascia lata on the medial side of the knee.

Lateral Aspect of the Knee Joint

The fibular collateral ligament is attached above to the lateral epicondyle of the femur and below to the head of the fibula. The tendon of the biceps is inserted into the head of the fibula, dividing into two parts to enclose the ligament. Beneath the fibular collateral ligament, the popliteus tendon is inserted into the lateral epicondyle of the femur. It is surrounded by a synovial prolongation from the knee joint. Behind the fibular collateral ligament, a capsular thickening extends from the lateral femoral condyle to the fibular head. This is known as the short fibular collateral ligament or arcuate ligament. The peroneal nerve lies just behind the biceps tendon and passes to the neck of the fibula.

Posterior Aspect of the Knee Joint

The popliteal fossa is a diamond-shaped space, which is bounded above by the hamstring muscles and below by the heads of the gastrocnemius The roof consists of the popliteal fascia, a continuation of the fascia lata. At the lower apex of the fossa, the posterior cutaneous nerve of the calf emerges through the fascia. It is traced upward where it originates as the uppermost branch of the posterior tibial nerve. Therefore, it may be used as a guide in dissecting through the popliteal fat. The popliteal vessels lie anterior and medial to the posterior tibial nerve. Lateral to the posterior cutaneous nerve of the calf is the external saphenous vein and the posterior cutaneous nerve of the thigh. Two nerves arise from the common peroneal nerve and pass over the lateral head of the gastrocnemius under the fascia lata. One is the lateral cutaneous nerve of the calf which supplies sensation to the anterolateral aspect of the proximal portion of the leg. The other is the

anastomotic peroneal branch which proceeds downward to join the posterior cutaneous nerve of the calf, forming the sural nerve. At the upper fossa, the common peroneal and the posterior tibial nerves join to form the sciatic nerve.

In making an incision over the popliteal space, the direction should be transverse to avoid keloid formation. Beneath the fascia, the posterior cutaneous nerve of the calf is the first structure to be encountered. Lateral to it is the external saphenous vein, and farther lateral are the anastomotic peroneal nerve and the lateral cutaneous nerve of the calf. The posterior cutaneous nerve of the calf is traced through the mass of fat to the posterior tibial nerve, which is followed to the sciatic. The large vessels lie anterior and medial to the posterior tibial nerve. In dissecting the common peroneal nerve downward, the anastomotic peroneal and lateral cutaneous nerve of the calf must be preserved.

To facilitate exposure of the floor of the popliteal fossa, the tendon of the medial head of the gastrocnemius is sectioned and turned over laterally, retracting the popliteal vessels and nerves. The geniculate vessels must be ligated and cut. Likewise, the lateral aspect may be exposed by severing the lateral head of the gastrocnemius. On the floor of the fossa, the insertion of the semimembranosus into the tibia and its fascial expansions to the oblique popliteal ligament, the popliteal fascia and the tibial collateral ligament are seen. A large bursa is found between the semimembranosus and the medial head of the gastrocnemius, which frequently communicates with the knee joint.

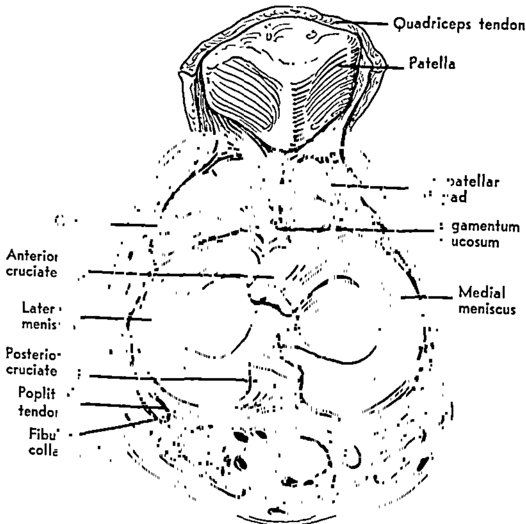


Fig. 405. Superior aspect of the tibia demonstrating the relationship of the menisci to other structures.

Semilunar Cartilages and Cruciate Ligaments

These will be discussed under their respective headings.

Movements of the Knee Joint

The knee joint is both a ginglymus (hinge joint) and a trochoid (pivot joint) articulation. It is capable of extension and flexion and, in certain positions, of internal and external rotation. During the first 20° of flexion, the movement is a rocking type of motion. After 20° of flexion, the movement is a gliding type of motion. It is beyond the first 20° that the supporting ligaments are relaxed, permitting not only gliding but also axial rotation. In the extended position, tautness of the ligaments prevents rotatory motion. At 90° of flexion, up to 40° of rotation is possible. Inward rotation is always greater than outward rotation. In complete flexion, rotation is decreased.

During flexion and extension, the menisci migrate slightly forward with extension and backward with flexion. The range of excursion of the medial meniscus is less than that of the lateral meniscus, the former being firmly anchored to the capsule and the tibial collateral ligament. The freedom of movement of the lateral meniscus makes it less likely to injury.

When extension is performed, the total area of the lateral femoral condyle and the corresponding surface of the medial condyle are expended. As movement is continued, the remaining extension movement occurs on the oblique portion of the medial condyle. This is accomplished by internal rotation of the femur until the remaining articular surface is used up. This is popularly known as the "screw-home" movement. The knee is locked in full extension, no axial rotation being possible The normal flexion-extension range is from 40° of flexion to 5° of hyperextension.

Lateral abduction and adduction motion is almost negligible in the fully extended position. In the flexed position, it varies from 6° to 12°.

Stability of the knee in the extended position is attained by the quadriceps anteriorly and the gastrocnemius and the popliteus posteriorly and tightening of the collateral ligaments. In the extended position, the center of gravity falls anterior to the center of knee joint motion, thereby aiding stabilization. The anterior aspects of the femoral condyles compress the lateral extensions of the infrapatellar fat pad.

SEMILUNAR CARTILAGES

The semilunar cartilages are two crescentic plates of fibrocartilage which are placed on the condylar surface of the tibia. They are triangular in cross section, the wide base facing externally, the apex directed internally. Each meniscus has two fibrous extremities or horns which are attached about the intercondylar area on the tibia. The semilunar cartilages function to deepen the surface upon which the femoral condyles roll and, by their mobility, to continue to accommodate the femoral condyles as the latter glide forward and backward. They act as shock absorbers to cushion a blow from above and below. Removal of a meniscus results in degeneration of the apposing cartilaginous articular surfaces. The outer aspect of the meniscus is convex, while the medial border is concave The superior surface is concave, and the inferior surface is flat.

The lateral semilunar cartilage is larger in breadth than the medial meniscus and forms an almost complete circle, the horns attaching adjacent to each other. The anterior horn is attached to the tibia just in front of the intercondylar eminence, lateral to and partly beneath the anterior cruciate ligament. The posterior horn is attached directly to the intercondylar eminence and sends a ligament (ligament of Wrisberg) to the posterior cruciate ligament and the medial femoral condyle. The cartilage is separated from the fibular collateral ligament by the capsule and the popliteus tendon.

The medial meniscus is semicircular and forms a segment of a much larger circle than the lateral. Its small anterior horn is attached to the intercondylar area in front of the anterior cruciate ligament. The larger posterior horn is fixed to the intercondylar area in front of the attachment of the posterior cruciate and behind the posterior horn of the lateral meniscus The outer aspect of the meniscus is attached to the posterior deep fibers of the tibial collateral ligament and capsule.

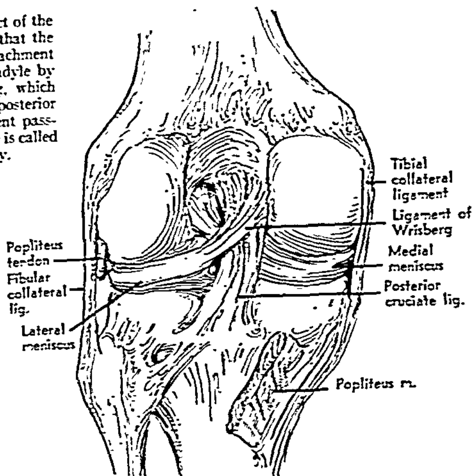


FIG. 406. Posterior aspect of the stripped knee joint. Note that the lateral meniscus gains attachment to the medial femoral condyle by the ligament of Wrisberg, which passes posterior to the posterior cruciate. A similar ligament passing anterior to the cruciate is called the ligament of Humphrey.

The anterior horns are connected by a fibrous band, the transverse ligament of the knee. The circumferential aspect of each cartilage is loosely attached to the rim of the tibia by the coronary ligament. The meniscus is avascular except at its peripheral zone of attachment.

MECHANISM OF INJURY

During flexion and extension of the knee joint, the menisci glide slightly posteriorly and anteriorly, respectively. In extreme flexion, the posterior half of the meniscus is compressed between the posterior aspects of the tibial and the femoral condyles. If, in the position of flexion, the femur is rotated internally, the posterior segment of the medial meniscus is forced toward the center of the joint by the medial femoral condyle. The posterior horn may be trapped in this position by sudden extension of the knee. Traction of the cartilage toward the center of the joint tears it from its peripheral attachment and causes a longitudinal splitting of its substance. The medial meniscus by being attached firmly to the deep posterior fibers of the tibial collateral ligament is more likely to be split than the more mobile lateral meniscus. This explains the frequency of tears in the medial meniscus which are confined, at least at its onset, to the posterior horn. The cartilage moves with the force of the femur rather than the tibia. Therefore, the beginning of the tear is on the inferior aspect to which it may be confined or extend to the superior surface. The larger the meniscus, the more susceptible it is to tearing. This, too, explains why the tear starts in the larger posterior horn.

The converse of the above mechanism, i.e., external rotation of the femur while the knee is flexed, will displace the posterior half of the lateral meniscus toward the center of the joint. Because the lateral meniscus is mobile, it cannot be torn longitudinally by the mediolateral distracting force such as exists on the medial side of the joint. Instead, during sudden extension of the knee, an anteroposterior distracting force tends to straighten the cartilage and imposes a strain on the medial concave rim which tears transversely or obliquely.

The tear rarely extends through the entire breadth of the meniscus.

In full extension, the anterior aspect of the tibial and the femoral condyles exert their compression forces against the lateral prolongations of the infrapatellar fat pad. Tear of the semilunar cartilages is rare in this position, unless it is part of an extensive injury involving disruption of the collateral and the cruciate ligaments and fracture of the tibial condyle. When the lateral or medial tibial condyle is depressed downward, the meniscus is often torn and displaced between the fragments.

The combination of flexion and extension superimposed upon internal or external rotation is necessary for injury to the menisci. Clinically, this is caused by the tibia's being fixed to the ground and unable to follow the violent torsional force of the femur. Internal rotation of the femur is a motion necessary to straightening up from the crouched or flexed position.

Clinicopathologic Correlation

The original tear occurs in the posterior segment and on its inferior aspect when the medial meniscus is involved. If the longitudinal tear involves the posterior third only, the entire meniscus by virtue of its elasticity springs back to its original position. If the tear extends forward beyond the level of the collateral ligament, the inner segment cannot be sprung back but instead is incarcerated between the medial femoral and the tibial condyles, and locking occurs. If the tear is extensive, the entire inner segment is completely displaced into the center of the joint, and no locking is possible. The displaced portion of the cartilage frequently undergoes hypertrophy.

Symptoms develop not because of the meniscal tear per se but because of tearing and hemorrhage in the peripheral attachments. The violent synovial contusion causes a synovial reaction in the form of swelling, congestion and marked effusion. Subsequent trauma to the mobile torn segment exerts less traction force upon the peripheral attachment with the result that the synovium reacts less intensely. This explains why each succeeding injury produces less severe symptoms than the previous one.

Because of lack of blood supply, a tear within the substance of the cartilage does not heal. If the tear is in the peripheral vascular zone, it heals by ingrowth of fibrous tissue from the synovium. When a tear within the cartilage extends to the periphery, only the peripheral portion heals.[2]

A transverse tear, such as occurs in the lateral meniscus, takes place at the junction of the anterior and the middle thirds. The tear begins on the concave edge and extends backward and laterally toward the convex margin. The lesion resembles a parrot beak. It is often associated with cystic degeneration of the lateral meniscus. Fibrous fixation of the cyst to the surrounding soft tissues incarcerates the lateral meniscus, thereby predisposing it to tearing. Because the fragment is small and the meniscus usually mobile, locking rarely occurs. Rarely does the transverse tear become complete.

When a cartilage is removed, it is replaced by a narrow, smooth, relatively inelastic structure closely resembling the original cartilage in appearance but composed of dense collagenous tissue. It is rarely torn.

Clinical Picture

The patient gives a history of a twisting injury to the knee while the joint was flexed. Usually, the leg is firmly fixed to the ground while the thigh is violently rotated. Most commonly, the leg, in relation to the thigh, is rotated externally and abducted. A football player is struck from the side. An acrobatic dancer performs a sudden turn or twist. The worker may rise erect from a squatting position in which the leg is placed sidewise with the foot pointed outward. Sudden severe pain is experienced, and he may feel "something tear" within the joint. Locking may not occur or may be momentary and reduced suddenly with a sensation of something slipping into place, usually by pulling or twisting the leg; or locking may persist and attempts to extend the joint accentuate the pain. Within a period of a few hours, marked effusion develops within the joint. Over ensuing days or weeks, during which time repeated efforts are made to extend the stubbornly locked joint, the degree of locking gradually lessens, and the

[2] King, D.: The healing of semilunar cartilages. J. Bone & Joint Surg. 18:333, 1936.

knee can be straightened almost fully or completely. This indicates forward extension of the tear.

Diagnosis

The following are the main diagnostic features:

Locking. The initial incident is not often associated with locking. The tear is confined to the region posterior to the coronal plane of the joint. Only when it extends into the anterior part of the meniscus at a subsequent injury does the displaced segment interpose between the tibia and the femur, preventing full extension of the joint. True locking occurs suddenly, and unlocking takes place with equal dramatic suddenness. Any gradual interference with full extension and gradual subsidence probably is due to other causes, such as effusion or hemarthrosis, hypertrophy or hemorrhage in the infrapatellar fat pad, fracture, or a tense popliteal cyst. Sudden locking and unlocking may also be caused by a loose body jammed between the articular surfaces.

Two conditions frequently simulate locking: (1) a hemorrhage between capsule and meniscus, either anterior or posterior to the collateral ligament, the result of a rotation strain, causes muscle spasm and interferes not only with extension but also with flexion; (2) contracture of the posterior capsule induced by prolonged rest in the flexed position.

Giving-Way. Buckling or giving-way of the knee joint when walking over uneven ground or when suddenly turning about suggests *a tear in the posterior segment of the meniscus.* The patient often relates that "something slips about in the joint." The joint feels insecure. Instability can also be caused by an old rupture of the anterior cruciate ligament or quadriceps insufficiency, but symptoms occur on descending stairs or jumping from a height.

Effusion is always present following the initial injury. It is due not to splitting of the meniscus but to tearing of the synovial or ligamentous attachments. If effusion is absent, an extra-articular lesion should be suspected. Subsequent injuries are characterized by a lessening degree of effusion until eventually, as the meniscus is easily displaced without damage to the peripheral attachments, synovial reaction is minimal or absent. Lesions of the lateral cartilage display a lesser amount of effusion than the medial, because its attachments are weak, and the synovium and the ligament are little affected.

Quadriceps Atrophy. Wasting of the quadriceps quickly follows any internal derangement of the knee. The vastus medialis is chiefly affected.

Condition of the Accessory Supporting Structures. One must evaluate laxity of the joint caused by recurring locking, effusion and giving-way. The presence of a torn collateral ligament and torn cruciate ligament should be established to determine whether or not reconstruction will be necessary. The unstable joint, particularly if the quadriceps is atrophied and weak, will delay convalescence and contribute to the development of traumatic osteoarthritis.

Tenderness. This may be present along the whole joint line. It indicates tearing or sprain of the peripheral attachments of the meniscus. Tenderness is most acute at one of the following points:

1. *Posterior peripheral attachment*—common, site of posterior segment
2. *Anterior peripheral attachment*—uncommon
3. *Collateral ligament*—most common and reliable for medial meniscus tears only. It indicates disruption of the deep fibers where it attaches to the meniscus. Tears of the ligament itself take place at the femoral condyle above the joint line.

Click. An audible click or snap is caused by the femoral condyle riding over an irregularity. This must not be confused with grating sounds caused by osteoarthritis, patellar clicking and snapping hamstring tendons where they run over the femoral condyle. Eliciting the click is especially important in establishing the presence of a lesion in the posterior segment of the meniscus which otherwise causes only vague symptoms, such as giving-way and little pain if any.

McMurray Test.[3] The patient is recumbent, and the knee is flexed until the heel touches the buttock. One hand steadies the knee, and the other hand grasps the heel. By twisting the leg into internal rotation, the posterior seg-

[3] McMurray, T. P.: The semilunar cartilages, Brit. J. Surg. 29:407, 1942.

ment of the lateral meniscus is tested, while external rotation tests the posterior segment of the medial meniscus. While the leg is forcefully rotated, the knee is gradually extended. The point in the arc at which a painful snap or click is felt or heard defines the site of injury in the meniscus. Thus, in full flexion the posterior segment is brought under pressure, and at a right angle, the middle segment. Beyond this point, no pressure can be exerted on the meniscus, so the method is of no value in testing for lesions of the anterior third. A lax meniscus without a lesion can produce a click but clinically is asymptomatic.

Lateral Meniscus

Injury of the lateral meniscus is only a little less common than that of the medial meniscus. It is characterized by less definite symptoms, a trivial type of accident, infrequency of locking, and production of little or no effusion The symptoms are often referred to the medial side of the joint. However, tenderness, pain on manipulation, and clicking occur on the lateral side.

Both Menisci

Repeated incidents of injury causing recurrent effusions result in marked laxity of capsule and ligaments plus quadriceps insufficiency. The anterior cruciate and the medial collateral ligaments may have been torn at the original injury. The resultant instability of the joint predisposes to injury of the lateral meniscus. Therefore, in internal derangement of long standing, lesions of both menisci should be suspected and sought after at operation.

Roentgenologic Findings. Shadows of the menisci can be outlined by using air as a con-



Fig. 407. Air arthrogram. The forceful adduction of the leg widens the lateral joint space, allowing the air to outline the lateral meniscus. The line drawn transversely designates the center of the joint for accurately lining up the central ray.



Fig. 408. Air arthrogram, outlining the lateral meniscus. The lateral position of the patella indicates that the anterior third of the meniscus is being visualized

trast medium. Only major tears and displacement of large segments can be defined. Lesser or atypical tears are difficult to see.

Technic. The skin is washed thoroughly with soap and water and then is painted with an antiseptic. Local anesthesia is employed. A needle is inserted laterally beneath the outer border of the patella. A 3-way stopcock is fitted to the hub of the needle, and a 50-cc syringe is attached All effusion is aspirated. Then air is injected in an amount sufficient to balloon the synovial pouch. The suprapatellar area bulges forward, and the patient complains of intense discomfort. The needle is withdrawn, and the puncture is sealed by pressure and collodion. Anteroposterior and lateral projection views are made and examined for other soft-tissue pathology. To outline the medial meniscus, the medial joint space is widened by a restraining bandage which pulls the thigh medially and another





Fig. 409. Air arthrogram. Normal exposure at left shows normal structures. Soft tissue technic (under exposure) at right brings out intrasynovial soft-tissue densities.

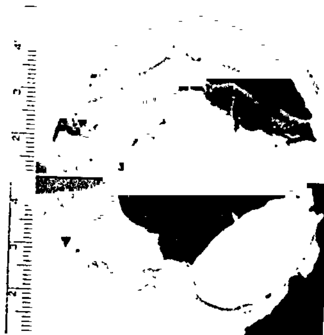


FIG. 410. Torn semilunar cartilage. The upper illustration shows the superior aspect, which does not appear too badly damaged. The true extent of loss of substance is seen on the inferior aspect shown in the lower illustration.

which pulls the leg laterally. In the direct anteroposterior projection, the middle third of the meniscus is outlined. By rotating the extremity externally or internally, the posterior and the anterior thirds, respectively, are brought into view. By reversing the lateral traction pull, the lateral joint space is widened, and the lateral meniscus silhouette is outlined. Finally, the needle is reinserted, and the air is allowed to escape.

Treatment. It is of prime importance that a locked knee be reduced within the first 24 hours. After this time, the effusion causes softening and loss of elasticity of the cartilage, which thereafter lacks the elasticity to spring back to its original position. Constant unsuccessful attempts at reduction only cause extension of the tear far anteriorly, the torn segment displaces more medially into the center of the joint, and almost full extension can be secured. This constitutes a false reduction. The anterior portion of the displaced segment may continue to interfere with the last few degrees of extension as demonstrated by a rubbery resistance toward the end of passive extension of the joint The loose fragment is displaced permanently.

TECHNIC OF MANIPULATION. In the treatment of a locked knee several methods can be used: (1) Longitudinal traction may be put on the leg. At the same time the leg is rotated externally and internally and is abducted and adducted. (2) The patient forcefully kicks the leg into extension while the surgeon at the same time pulls and rotates the leg internally. (3) The knee is fully flexed. If the medial meniscus is affected, the leg is forcefully rotated internally, and the knee is forcefully extended. For the lateral meniscus, the maneuver is performed with the leg rotated externally.

After reduction, the torn tissues should be allowed time to heal. A compression bandage is applied, and quadriceps exercises are instituted immediately. Then surgical excision must be done. A torn meniscus never heals unless it is torn from its peripheral attachment, but this latter type of lesion is extremely rare and cannot be identified clinically. A torn meniscus will cause recurring attacks, joint instability, predisposition to injury of the contralateral meniscus, and eventually a traumatic osteoarthritis. The entire meniscus is excised. A retained posterior segment will cause a giving-way of the knee when walking on rough ground and a sensation of something slipping about in the joint.

TECHNIC OF OPERATION. After the skin has been prepared aseptically, a tourniquet is applied. Next, the patient is placed with the knee flexed over the end of the operating table. The author prefers the Cave incision, which gives wide exposure. A transverse incision is made over the joint interval. It may be extended proximally at the posterior end and distally at the anterior end. The capsule is cut in the line of the skin incision in front and behind the collateral ligament. On the medial side, the sartorius tendon falls away posteriorly out of harm's way. On the lateral side, the popliteus tendon is seen over the posterior third as it ascends toward the femoral condyle. The synovium is incised, and the joint interval is widened by abducting or adducting the leg as the case may be. The coronal ligament which attaches the circumference of the meniscus to the rim of the tibia is seen and cut in its anterior third, and the cartilage is followed to its anterior attachment. The latter is severed, and the mobilized seg-

ment is passed posteriorly beneath the collateral ligament; the cartilage is freed backward to its posterior attachment, which is cut under direct vision. The interior of the joint is inspected for other pathology. The synovium and the capsule are sutured as one. The skin is closed, and a compression bandage is applied

Postoperative Treatment. On the second or the third day, quadriceps exercises are started and graduated so that several pounds of weight can be lifted with ease. Abstinence from weight-bearing for 3 weeks or more is required until the new meniscus is fully formed. Exercises are continued until muscle tone is adequate. Recurrence of effusion is a sign that exercises are excessive and should be discontinued temporarily.

Hemarthrosis most commonly follows removal of the lateral meniscus because of the proximity of the lateral geniculate vessels which frequently are severed. The compression bandage should not be removed too soon. The presence of blood distending the joint causes severe pain, swelling, and increase of local temperature. If allowed to remain within the joint, adhesions may form, restricting motion and making for a prolonged convalescence. The blood should be aspirated, and the compression bandage reapplied.

The infrapatellar branch of the saphenous nerve may be severed in the approach to the medial meniscus. It causes temporary anesthesia over a small area below the patella. Occasionally, the nerve may be incarcerated in the operative scar, causing pain on movement of the joint. If it cannot be freed by injecting locally a solution of local anesthetic under force, surgical neurolysis is necessary.

Extra-articular Displacement of the Medial Meniscus[4]

The coronary ligaments by which the medial meniscus is attached to the rim of the tibia prevent outward displacement of the cartilage during weight-bearing. When these ligaments are torn, tibiofemoral compression forces the cartilage out of the joint interval. The loose meniscal segment bulges outward beneath the tibial collateral ligament and, when excessively mobile, may migrate upward between femur and ligament or downward between tibia and ligament. Occasionally, it may protrude anterior or posterior to the ligament.

Extra-articular displacement produces an ill-defined clinical picture, and often diagnosis is made on surgical exploration. Because the meniscal defect is such that tibiofemoral movement is not seriously interfered with, pain is moderate, locking never occurs, and effusion is minimal. Pain and tenderness over the inner aspect of the knee, particularly at the center or the upper or the lower attachments of the medial collateral ligament, suggest strain of this ligament. However, when conservative treatment fails to relieve symptoms, displacement of the meniscus is generally the cause.

Clinical Picture. The onset may be insidious or sudden. Pain occurs on weight-bearing and is aggravated at a certain arc of flexion corresponding to the portion of cartilage involved. Pain is often initiated or aggravated by external rotation of the leg. Swelling is absent or minimal. Locking does not occur. Frequently, no history of injury can be obtained.

Examination reveals tenderness localized to the center of the tibial collateral ligament or at its upper or lower attachment. Tibial collateral pain is produced inconstantly when abducting the leg upon the thigh. Apparently, the symptom is produced only when the cartilage is displaced beneath the ligament. Pain is also reproduced by external rotation of the tibia with the knee flexed to 90°.

Operative Findings. The middle third of the medial meniscus is thickened peripherally and thinned at its inner border. It appears as though the cartilage mushrooms out of the joint space and bulges the ligament outward. This external protrusion is more prominent when the tibia is forcefully adducted medially upon the femur. Occasionally, the cartilage is completely herniated out of the joint and is displaced upward between the ligament and the femur, or downward between the ligament and the tibia. Other pathology may be present in addition to the herniation, such as a healed tear or a meniscal cyst. Meniscectomy cures the condition.

[4] Smith, F. B., and Blair, H. C.: Tibial collateral ligament strain due to occult derangements of medial meniscus, J. Bone & Joint Surg 36A 85, 1954.

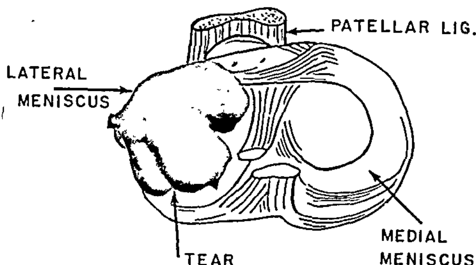


Fig. 411. Discoid cartilage. The actual specimen is superimposed upon a drawing of the superior articular surface.

CONGENITAL DISCOID MENISCI

At birth, the menisci are thick, flat, oval-shaped, cartilaginous masses which cover almost the entire articular surface of the tibia. The central portion of each meniscus gradually absorbs until the cartilage assumes its normal adult configuration. If normal resorption does not occur or is incomplete, the meniscus retains its fetal discoid appearance. The condition is more common on the lateral side of the knee joint.

Pathology

Often the discoid meniscus is thicker peripherally, while the central portion is thin and transparent. At birth the surface is smooth, but with weight-bearing a depression forms anteriorly and another posteriorly to accommodate the femoral condyle in extension and flexion, respectively. Between the depressions, a ridge develops which is responsible for the characteristic snapping sound as the femoral condyle rides over it. The discoid meniscus is subject to multiple tears and cystic degeneration.

Clinical Picture

At or shortly after birth, the mother notices an audible snap associated with movement of the knee. On examination, motion of the joint is interrupted temporarily at about 160°, a loud and palpable snap or clunk is apparent, and beyond this point movement proceeds smoothly. No synovial effusion is present, and no tenderness is elicited With passage of time over many months or years, the snap and jerking motion gradually disappears, signifying that the cartilaginous ridge is worn down Occasionally, the annoying symptom persists into adulthood. The symptoms and the findings of a tear and cyst formation are frequently superimposed.

Treatment

In infancy, no treatment is required, as the cartilage may be worn down by the pressure of weight-bearing. If the meniscus is torn or



Fig. 412. Cyst of the lateral semilunar cartilage. The extent of the tumor indicates stretching and attenuation of the lateral collateral ligament of the knee. Reconstruction of the ligament may be required.

develops a cyst, it should be removed. Cyst formation about a meniscus should suggest a discoid cartilage.

Cysts of the Semilunar Cartilage.[5] Cysts varying from multiple small cysts to one large plum-sized cyst may develop within or just external and attached to a meniscus, either medial or lateral. The very minute cysts generally are multiple; they occur in clusters and are situated within the substance of the cartilage. The single large cyst appears as a tensely distended sac, multiloculated, bluish in color, surrounded and engulfed by reactionary fibrous tissue; it contains clear gelatinous fluid and is attached to the lateral aspect of the meniscus, usually the lateral one and commonly in its middle or anterior third. Microscopically, the sac cavity may extend within the substance of the cartilage. Frequently, evidence of previous trauma to the meniscus, such as an incomplete transverse or longitudinal tear at the site of cyst attachment, suggests traumatic degeneration as the etiology. The large parameniscal cyst may remain entirely within the capsule, but usually it stretches the capsule and may distend a portion of the collateral ligament and may even become entirely subcutaneous.

Clinically, the patient is a young male, frequently athletic, between 20 and 30 years of age If the meniscus had been damaged previously, a history of instability or buckling associated with pain and repeated effusion may be obtained. With development of the cyst, typically there develops a dull constant ache over the involved meniscus which is aggravated by increased activity. The swelling may be apparent generally over the lateral side of the joint and is firm and tender to palpation. It is largest while the knee is fully extended and recedes somewhat during flexion. When the cyst projects through the collateral ligament, limitation of extension and occasionally of flexion results.

The condition must be differentiated from a pedunculated tag of meniscus or an entire segment of meniscus which displaces laterally beyond the confines of the joint to produce a lateral swelling. However, the pathology is apparent on operative exposure and treatment is the same. A bursa beneath the lateral ligament or a true outpouching of the synovium, a loose body in the lateral joint compartment, and an exostosis are other conditions producing localized lateral swelling.

Treatment consists of removal of the cyst and the meniscus. A transverse incision is made directly over the joint line over the extent of the meniscus. The posterior extremity of the incision may be extended upward and the anterior extremity downward if more exposure is desired. The collateral ligament is identified and avoided if possible as the cyst is dissected free of cicatrix. The meniscus is exposed by deepening the incision in front of and behind the collateral ligament; its anterior, posterior and capsular attachments are cut; and the meniscus with its attached cyst is removed.

CALCIFICATION AND OSSIFICATION OF MENISCI[6]

Cartilage in any location may become calcified with advancing age. This often constitutes an intermediate stage in eventual ossification. Most frequently, this occurs in the chondral portions of ribs but may also take place rarely in the semilunar cartilage. Calcium may also be deposited in degenerated and necrotic tissue. Thus, a posterior horn of a meniscus, repeatedly traumatized, degenerates and provides a suitable locale for deposition of calcium salts. Therefore, the following two types may be distinguished:

1. **Primary Type.** This occurs in patients of advanced age, usually afflicted with degenerative arthritis. Calcium is deposited throughout both cartilages, frequently in both knees. Radiologically, the calcified menisci are revealed as transversely disposed radiopaque white lines in the tibiofemoral interval. The symptoms are those of degenerative arthritis. With passage of time, ossification develops, and actual trabeculations are demonstrable. The menisci do not require removal.

2. **Secondary Type.** The younger individual with symptoms of a torn meniscus is affected. A vague deposit is observed in the region of the posterior horn and may be confused with a loose body. It is considered merely as a

[5] Smillie, I. S: Injuries of the Knee Joint, Edinburgh, Livingstone, 1946.

[6] Cave, E. F.: Calcification of the menisci, J. Bone & Joint Surg. 25.53, 1943.

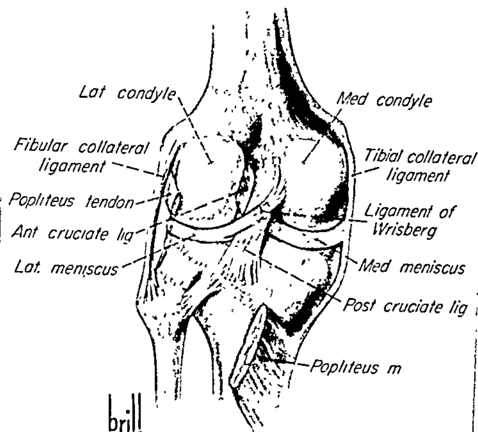


FIG. 413. Posterior aspect of the stripped knee joint. Observe the relation of the posterior cruciate ligament to the medial meniscus, the anterior cruciate ligament to the medial meniscus, the posterior cruciate ligament to the ligament of Wrisberg (DePalma, A. F.: Diseases of the Knee, Philadelphia, Lippincott)

manifestation of a torn meniscus which requires removal.

LIGAMENTS OF THE KNEE

The ligaments of the knee are composed of the medial and the lateral collateral ligaments, the cruciates and the capsule. Their function of stabilizing the joint is passive and secondary to active muscle tone and control. Therefore, they are more likely to be ruptured when the muscles are relaxed or weak.

CRUCIATE LIGAMENTS

The anterior cruciate ligament is attached anteriorly to the nonarticulating portion of the upper tibial surface in front of the intercondyloid eminence at the anterior attachment of the lateral meniscus It is directed upward and backward to attach to the posterior aspect of the inner surface of the lateral condyle of the femur. It is invested by synovium. It functions to prevent anterior displacement and controls rotation of the tibia on the femur. It is tense only in hyperextension and in acute flexion. Behind the anterior cruciate is located the posterior cruciate ligament. It is attached to the posterior intercondyloid fossa of the tibia just behind the posterior horn of the medial meniscus. It is directed upward, forward and medially, crossing the anterior cruciate to be attached to the lateral surface of the medial femoral condyle. It may blend with the posterior capsule. The posterior horn of the lateral meniscus sends a ligament to the posterior cruciate (ligament of Wrisberg), then passes posterior to the latter to attach to the medial femoral condyle Occasionally, a similar ligament passes anterior to the cruciate (ligament of Humphry). The function of the posterior cruciate is to prevent posterior displacement of the tibia on

the femur. Some portion of the ligament is tense throughout flexion and extension.

In the fully extended knee, both the cruciates and collateral ligaments act to restrain anteroposterior movements of the tibia on the femur. In the flexed position the collateral ligaments, particularly the lateral one, relax and only the cruciates control anteroposterior displacement. Therefore, when the anterior cruciate alone is interrupted, and the fully extended knee is examined, only a slight anterior displacement of the tibia on the femur (2 to 5 mm.) can be effected. As the knee is flexed, collateral ligament control is lessened and anterior displacement becomes exaggerated to as much as 2 cm. If the anterior cruciate and the tibial collateral ligaments are cut, anterior displacement is considerable (anterior drawer sign) even in the fully extended knee. It is apparent that the tibial collateral ligament, in extension, is a strong checkrein to anteroposterior excursions. When all ligaments are divided the joint is extremely unstable. Clinically, a very positive drawer sign and abnormal widening of the medial joint interval on abducting the leg indicates rupture of the tibial collateral and the anterior or both cruciate ligaments.

Anterior Cruciate Ligament. This ligament is attached anteriorly to the anterior aspect of the tibial spine at the anterior insertion of the lateral meniscus. It is directed upward, backward and laterally to attach to the posterior part of the medial surface of the lateral femoral condyle. The ligament is tense throughout the range of motion and at the extremes of flexion and extension of the joint. Its main functions include control of:

1. Forward gliding of the tibia on the femur (main function)
2. Lateral mobility in flexion and extension
3. Rotation in flexion and extension
4. Hyperextension
5. Hyperflexion

Therefore, it is stretched or ruptured by a force driving the femur backward while the knee is flexed and the tibia is fixed, by hyperextension, by violent rotation, by abduction (accompanied by rupture of the medial collateral ligament and occasionally a depressed fracture of the lateral tibial condyle), and by dislocation of the knee. It is frequently associated with rupture of the medial collateral ligament. Abduction and rotation injuries produce, in addition, meniscal tears.

Rupture takes place at one of three points:

1. *Inferior insertion*—tibial spine may be avulsed. Ligament is intact.
2. *Superior insertion*—ligament is frayed.
3. *Throughout its extent*—fibers within the synovial covering are stretched and torn. Ligament externally appears to be intact. Usually follows abduction type of injury.

Clinical Picture of Recent Rupture. A history of violent trauma is often obtained. A hemarthrosis is invariably present and causes marked swelling, severe pain, generalized joint tenderness and systemic reaction. Although a torn medial meniscus and a ruptured medial collateral ligament are frequently associated, a locked knee precludes the presence of a torn cruciate. Conversely, a torn cruciate permits widening of the tibiofemoral interval and allows the meniscus to spring back into place.

Drawer Sign. Muscle spasm and hemarthrosis interfere with flexing the joint and testing for forward gliding of the tibia. Preferably under general anesthesia, the joint is aspirated. The presence of fat globules in the bloody fluid suggests a fracture, either an avulsion of the tibial spine or a depressed fracture of the lateral tibial plateau. The joint is flexed, and by placing both hands behind the upper leg the tibia is drawn forward. Anterior mobility is excessive when rupture of the medial collateral ligament is associated.

Treatment. It is possible that a cruciate ligament tear can heal by prolonged immobilization. If continuity is not restored, it is well compensated by strong muscle control, especially by the quadriceps apparatus. A well-developed quadriceps will provide sufficient stability for ordinary activity, as walking and standing, but not for rigorous occupations or athletics. The question of whether treatment shall be conservative or surgical depends upon these factors and whether or not associated surgical lesions are present, as a torn meniscus and a ruptured medial collateral ligament.

A severe injury often results in a triad, a tear of the medial meniscus, rupture of the medial collateral ligament and rupture of the anterior cruciate. Hemarthrosis and muscle

spasm make it almost impossible to determine the extent of damage. It is imperative to ascertain immediately whether or not the cruciate has been torn. Delay permits the ligament to retract, and repair is difficult. Under anesthesia, the joint is aspirated, and the drawer sign elicited.

Conservative Treatment. The joint is aspirated, and a compression bandage is applied. A posterior molded plaster splint is applied with the knee in extension. When swelling has subsided, a skin-tight cylinder cast is applied. Quadriceps exercises are started hourly with progressive increase in weights. After removal of the cast 12 weeks later, quadriceps exercises are continued several months longer.

Surgical Treatment. Reconstruction of a recently torn anterior cruciate ligament generally is one phase of a procedure involving repair of the medial collateral ligament and removal of the medial meniscus. The technic varies, depending upon the site of the tear.

Technic for Reattaching Superior Insertion. A strong silk suture is crisscrossed through the proximal portion of the distal segment of ligament, two strands emerging from the proximal end. A ⅜ inch drill hole is made in the lateral femoral condyle from without inward and emerges at the normal attachment on the lateral wall of the intercondylar notch. The silk strands are threaded through the hole and are drawn out externally; then they are drawn taut and tied over a bone peg. Instead of using a bone peg, a second hole is drilled through the external cortex adjacent to the first hole; one silk strand is passed through it and tied to the other strand over the intervening cortex.

Technic for Reattaching Inferior Insertion. A silk suture is crisscrossed through the distal portion of the proximal segment of ligament, two strands emerging through the distal end. A ⅜ inch drill hole is made from without inward, starting at the upper anteromedial aspect of the tibia and emerging within the joint at the inferior point of attachment of the

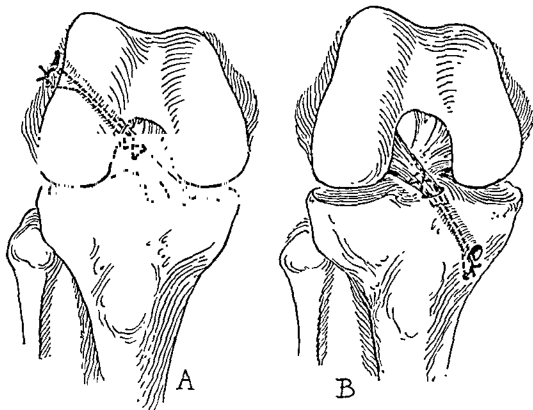


FIG. 414. Repair of the anterior cruciate ligament. When the ligament is separated at its proximal attachment, the drill hole is made so as to enter the intercondylar fossa at its posterior aspect. To restore the anterior attachment. the drill hole enters the joint at the anterior end of the tibial spine.

anterior cruciate. The strands are threaded through the hole, drawn up taut and tied over a bone peg. Or one strand may be threaded through a superficially drilled adjacent hole and the silk strands tied over the intervening bridge of cortex.

The technic for reconstructing an entire ligament is described under "Old Tears of the Anterior Cruciate Ligament."

FRACTURE OF THE TIBIAL SPINE. In youth, the anterior cruciate ligament is strong and, instead of rupturing at the anterior insertion, the bone is avulsed. The common mechanism is a force which drives the femur backward while the tibia is fixed and the knee is flexed. Rupture of the medial collateral ligament is rarely associated.

Clinically, a hemarthrosis is invariably present, and the aspirated fluid contains fat globules. Abnormal anteroposterior mobility is demonstrable under anesthesia. The x-ray film reveals the fragment which varies in size and displacement.

Treatment consists of attempted reduction by hyperextension followed by immobilization. A large fragment is easier to reduce, because it is compressed into place by the femoral condyles. If the fragment is greatly displaced, constituting in effect lengthening of the anterior cruciate, it must be replaced and fixed by the method described for reattaching an inferior insertion of the anterior cruciate. If the injury is of several weeks' duration, the fragment becomes covered by synovial membrane and is difficult to find. The synovium over the ligament should be incised and reflected In addition, the crater in the tibial plateau may be filled with callus and must be gouged out before the tibial spine can be reapproximated.

OLD RUPTURE OF THE ANTERIOR CRUCIATE LIGAMENT. A knee with a strong quadriceps, an intact medial collateral ligament, and no laxity of the capsule will display good function even in the presence of a ruptured anterior cruciate. A lesion of a meniscus with recurring incidents of locking effects atrophy and weakness of the quadriceps and laxity of all ligamentous structures. Marked instability of the knee and excessive anterior mobility of the tibia on the femur result, eventuating in a degenerative arthritis. Removal of the torn meniscus or menisci and repair of the collateral ligament followed by redevelopment of the quadriceps generally provides a satisfactory joint, although the cruciate is interrupted. Reconstruction of the cruciate ligament is rarely indicated. Relative indications include marked instability, especially when descending stairs (insufficient quadriceps compensation), and the demands of a vigorous occupation and athletic activities.

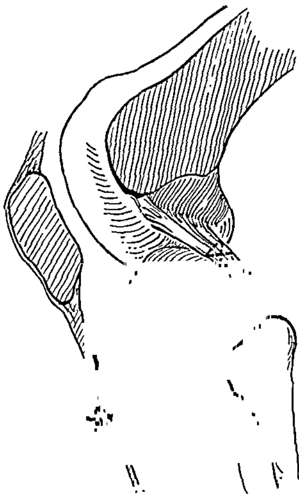


FIG. 415. Repair of rupture of the posterior cruciate at the inferior insertion. The suture passes through a drill hole which enters the joint at its posterior aspect.

Clinically, a history is obtained of an initial injury followed by recurring incidents of giving-way or locking, effusions and instability, particularly for descending stairs. The quadriceps is atrophied, and a positive anterior drawer sign is elicited. With the knee in flexion, excessive medial rotation of the tibia on the femur is noted. (Normally, the anterior cruciate wraps around in front of the posterior cruciate, restricting internal rotation.) Excessive external rotation, if present,

is due to laxity of the medial collateral ligament and capsule. (The anterior cruciate unwinds in external rotation.) Signs of meniscal and medial collateral ligament injury should be sought. The x-ray film is negative, except possibly for traumatic osteoarthritis.

Conservative Treatment. In the absence of meniscal and collateral ligament injury, redevelopment of the quadriceps is sufficient.

Surgical Treatment. Insufficient ligamentous structure remains. Replacement of the anterior cruciate is accomplished by utilizing a strip of fascia lata, as described by Hey Groves[7] The technic is described in the following section.

Posterior Cruciate Ligament. This ligament is attached to the posterior part of the tibial plateau behind the tibial spine. Its fibers pass upward, forward and medially, behind the anterior cruciate ligament, to attach on the anterior portion of the lateral surface of the medial femoral condyle. Its fibers are taut, both in flexion and extension, and it assists in preventing hyperextension and hyperflexion. Its main function is to prevent backward displacement of the tibia on the femur.

Rupture of the posterior cruciate ligament takes place as part of generalized destruction of ligaments such as follows a dislocation. It results from a force which drives the tibia backward while the knee is in flexion, e.g., a dashboard injury. Rupture takes place at either end of the ligament. In young individuals, avulsion of bone from the posterior rim of the tibia may occur instead of rupture at this insertion. Excessive backward mobility of the tibia, a "posterior drawer sign," is pathognomonic.

TREATMENT. Recent rupture of the posterior cruciate, being part of extensive ligament and meniscal damage, may be repaired at the same surgical procedure.

Technic for Reattaching Inferior Insertion. Through a popliteal approach, the capsule is opened. A crisscross suture is placed through the ligament. A ⅜ inch hole is drilled from the point of insertion of the ligament downward and forward to emerge on the anteromedial aspect of the tibia. The 2 silk strands are drawn through, pulled up tight and tied over a bone peg. If a large fragment has been avulsed, it is replaced in the tibial defect and fixed with a screw.

Technic for Reattaching Superior Insertion. Through an anterior approach a crisscross suture of strong braided silk is placed through the loose end of ligament. The strands are threaded through a hole made from without inward in the medial femoral condyle, emerging at the anterior aspect of the lateral surface. The threads are drawn through, pulled up taut and tied over a bone peg or by running one thread out through an adjacent drill hole.

Old rupture of the posterior cruciate existing as an independent lesion is rare. It causes severe instability which can be lessened by quadriceps strengthening. More commonly, it is associated with an old rupture of the anterior cruciate and the medial collateral ligaments and meniscal lesions.

Repair of Old Ruptures of the Anterior and the Posterior Cruciate Ligaments.[8] A longitudinal incision is made over the lateral aspect of the knee and is extended upward to the mid-thigh. Two strips of fascia lata, each 1 inch wide, are dissected downward, one as far as the lateral femoral condyle, the other to the lateral tibial condyle.

The knee joint is exposed by a parapatellar incision. This may be extended upward into the quadriceps and the patella displaced laterally for excellent exposure. Three drill holes, each 13/32 inch in diameter, are made for passage of the new ligaments. One is drilled in the medial condyle of the femur, from without inward, extending from near the medial epicondyle to a point anteriorly on the lateral wall. The second hole is drilled from the anteromedial aspect of the tibia externally to a point in front of the tibial spine internally. The third hole is drilled transversely through the posterior portion of the lateral condyle of the femur.

The fascia lata strips are twisted. To replace the anterior cruciate, one strip is drawn through the hole in the lateral femoral condyle; then it is passed through the hole in front of the tibial spine and is brought out

[7] Hey Groves, E. W.: Operation for repair of the cruciate ligaments, Lancet 2.674, 1917.

[8] Cubbins, W R., Callahan, J. J., and Scuderi, C. S: Cruciate ligament injuries, Surg, Gynec & Obst. 64:218, 1937.

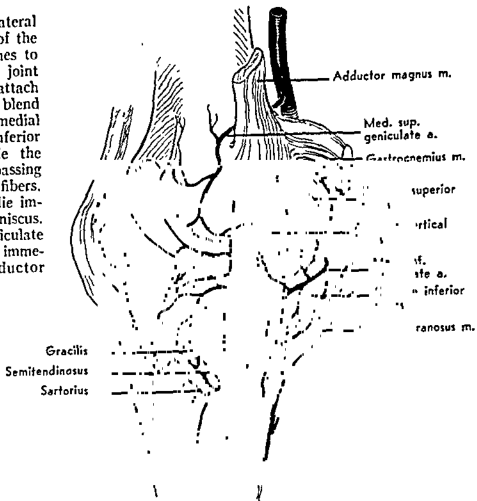


Fig. 416. The tibial collateral ligament. The lower end of the vertical component attaches to the tibia well below the joint line. The oblique fibers attach to the joint margins and blend with the periphery of the medial meniscus. The medial inferior geniculate vessels overlie the oblique fibers before passing beneath the long vertical fibers. Note that these vessels lie immediately below the meniscus. The medial superior geniculate vessels enter the joint immediately above the adductor tubercle.

and fixed to an osteoperiosteal bed on the anteromedial surface of the tibia The second fascial strip is used to replace the posterior cruciate. It is passed backward above the head of the fibula between the biceps tendon laterally and the lateral tibial condyle medially. (It must not be made to encircle the fibular head or the biceps tendon unless the peroneal nerve is first isolated and retracted.) Next, the strip is inserted beneath the lateral head of the gastrocnemius, then through the capsule into the knee joint. It is passed through the hole in the medial condyle of the femur. To this point, the posterior cruciate is reconstituted. It emerges externally near the medial epicondyle and, by extending it downward to be fixed to the inner side of the medial tibial condyle, the medial collateral ligament is reconstructed.

Postoperatively, the knee is immobilized in full extension for 6 weeks. Quadriceps setting exercises start within a few days after operation. After removal of the cast, a support must be worn for weight-bearing until quadriceps power is well developed.

Collateral Ligament

Medial (Tibial). This ligament is an extensive structure which attaches above to the medial femoral condyle adjacent to the adductor tubercle and below to the medial tibial condyle. Its deep fibers give attachment to the medial meniscus. It is taut only in extension and limits abduction, rotation and forward gliding of the tibia on the femur. Partial rupture or sprain occurs usually at its femoral attachment. Complete interruption in continuity can take place at any point or throughout its entire length.

Partial Rupture or Sprain. Partial avulsion of the long anterior fibers from their attachment to the medial femoral condyle is a very frequent injury, occurring as a result of a minor twisting strain with the knee in flexion. It causes severe pain over the medial aspect of the joint. On examination, there are

swelling and tenderness over the medial femoral condyle, the maximum tenderness being well localized to the point of attachment of the ligament. The pain is accentuated by abducting the leg while the knee is fully extended, but no widening of the joint interval occurs. External rotation of the leg will also reproduce the pain.

Treatment consists of injecting the area with a local anesthetic, puncturing the site of maximum tenderness with a large-bore needle and applying an elastic bandage. Immediate weight-bearing activity is permitted. Good results have been reported from the injection of Hydrocortone. Symptoms subside within a few days to several weeks.

Post-traumatic Para-articular Ossification (Pellegrini-Stieda's Disease). A small proportion of cases of sprain, instead of improving, grow progressively worse. Pain becomes more intense, and active flexion is restricted to as little as 40°.

As a result of tearing and shredding of fibers at their femoral attachment, or a severe blow to this site, a hematoma and inflammatory edema develop. The damaged soft tissues become degenerate and necrotic, forming an ideal environment for deposition of calcium salts. The inflammatory swelling with its calcium deposits interfere with backward gliding of the ligament during flexion of the knee joint. Eventually, the inflammation subsides with partial or complete resorption of the calcium salts. Or the mass becomes ossified and occasionally connected by a pedicle to the femoral condyle. A bony mass constitutes a permanent obstruction to movement of the ligament. Adhesions form between the ligament and the surface of the condyle below the epicondyle, with further restriction of movement.

Clinical Picture. The condition should be suspected when symptoms of a single sprain fail to subside. Instead, after several weeks, pain becomes worse, and now are added weakness and limitation of flexion. Examination reveals, in addition to tenderness, a diffuse indurated swelling in the soft tissues about the femoral condyle Restriction of flexion is characteristic. Attempts to flex the joint passively cause severe pain over the upper ligamentous attachment and a peculiar elastic resistance. The intensity of symptoms gradually lessen until, after several months to a year, pain is minimal but loss of flexion persists. At this time a bony prominence is palpable. It may be fixed to the underlying bone. The quadriceps is atrophied and weak.

Roentgenologic Findings. At about the third or the fourth week after injury, a small, narrow, elongated amorphous shadow forms adjacent to the medial aspect of the femoral condyle. The hazy, radiopaque, ill-defined shadow may increase in size, even assuming large proportions, particularly following ill-advised massage and forcible manipulation of the joint. With resorption, the shadow reduces in size and becomes increasingly dense and well-defined as ossification takes place. A bony projection, sometimes attached at its base to the femoral condyle, points distally. Occasionally, the shadow of calcium salts will disappear without subsequent bone formation.

Treatment. An ordinary sprain treated by needling and elastic compression should prevent calcified deposits. Once calcium deposits are established, injection of a local anesthetic followed by multiple punctures frequently effects rapid resorption of the calcium and subsidence of symptoms. Massage and passive movement must be avoided during the acute stage, as massive calcification may be encouraged. For the same reason, surgical removal of a bony mass is contraindicated.

When the condition becomes quiescent, usually after 9 months to a year, surgical treatment is directed toward restoring flexion. The bony mass is excised, and the joint is fully flexed to rupture adhesions.

Complete Rupture. This injury takes place as a result of violent abduction while the knee is fully extended. Often it is part of extensive damage to the anterior cruciate ligament and medial meniscus and fracture of the lateral tibial condyle.

Clinically, the joint is markedly distended with blood. Maximum pain and swelling occur over the medial aspect. Under anesthesia, a wide gap in the medial joint interval is demonstrated by abduction in full extension

Treatment. The ligament can heal with prolonged immobilization, although with some degree of lengthening. In the presence of a strong quadriceps, the result is satisfactory. However, complete rupture of the medial collateral ligament rarely occurs as an isolated

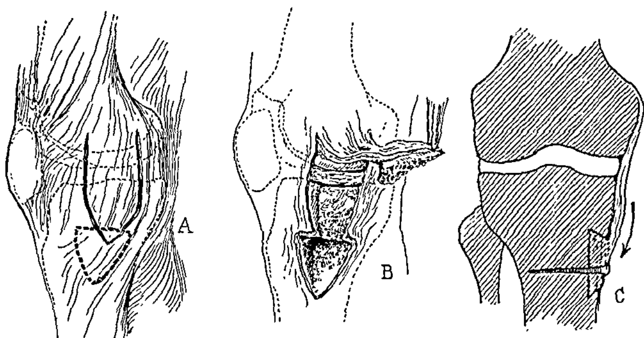


Fig. 417. Old torn and lax tibial collateral ligament of the knee, surgical correction. (Mauck's procedure)

lesion. The anterior cruciate is almost always involved. Therefore, maximum stability will be provided by surgical repair followed by quadriceps strengthening exercises.

Tears of the collateral ligament occur at any point, and they assume many bizarre forms. A flap of ligament may be interposed in the joint between the condyles. If direct suture is impossible, it may be reconstructed by using strips of fascia lata or an adjacent hamstring tendon.

Old Rupture. After the ligament heals by the interposition of fibrous tissue, it is elongated, particularly so when immobilization has been inadequate. It produces instability which is accentuated by loss of the anterior cruciate and a weak quadriceps Therefore, the effect of quadriceps redevelopment must first be ascertained. If lateral instability persists, reconstruction of the ligament is indicated One must be certain that the giving-way incidents are due to a lax ligament rather than a meniscus lesion. The latter in itself would retard improvement of the quadriceps.

Technic.[9] This procedure is suitable when the medial collateral ligament is intact but lax. A longitudinal incision is made over the medial aspect of the knee. Two parallel incisions are made in the capsule, one on each side of the ligament. They are extended distally about 6 cm. beyond the rim of the tibia where a flap of bone is elevated with the ligament. By raising the flap, the medial meniscus may be removed through the opening. Next, the ligament with attached bone is drawn as far distally as possible. At this point, a notch is made in the tibia into which the bone flap is fixed with a screw. Postoperatively, the knee is immobilized for 6 weeks, but quadriceps setting exercises are started almost immediately.

When the ligament has been thoroughly destroyed, it may be replaced by one of the hamstrings, preferably the semitendinosus.[10] The semitendinosus tendon is severed at the level of the femoral condyle. The proximal end of the distal segment is fixed to a groove cut in both the femoral and the tibial condyles at the site of attachment of the medial ligament. Immobilization for approximately 6 weeks is required.

Lateral (Fibular). This ligament of the knee joint is a strong, rounded band which

[9] Mauck, H. P.: A new operative procedure for instability of the knee, J. Bone & Joint Surg 18:984, 1936

[10] McMurray, T. P., Operative treatment of the internal lateral ligament of the knee, Brit. J. Surg 8:267, 1920.

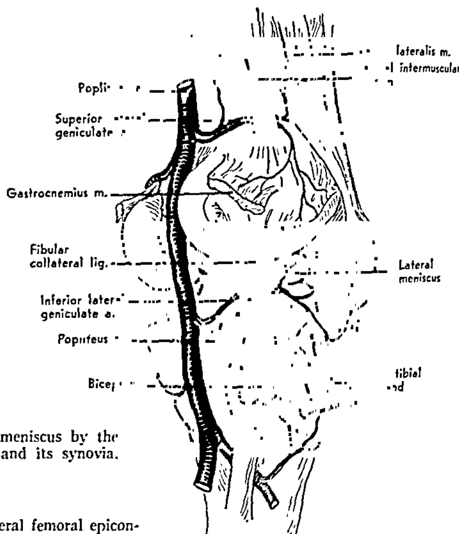


FIG. 418. The fibular collateral ligament. The superior lateral geniculate vessels enter the joint above the lateral epicondyle and immediately penetrate the substance of the vastus lateralis. The inferior lateral geniculate vessels usually lie higher than those depicted here; often they are in close contact with the lateral meniscus. It anastomoses with the recurrent vessel from the anterior tibial. The fibular collateral ligament attaches above to the lateral epicondyle of the femur and below to the head of the fibula. It is separated from the periphery of the lateral meniscus by the tendon of the popliteus and its synovia. sheath.

attaches above to the lateral femoral epicondyle and below to the head of the fibula. It has no attachment to the lateral meniscus, being separated from the latter by the popliteal tendon. With the knee flexed, the ligament is relaxed and unaffected by rotation strains. It is rarely injured and then only by violent adduction of the leg and when the knee is completely dislocated Tearing of the common peroneal nerve is often associated, the combination of the two injuries being known as "the ligamentous-peroneal nerve syndrome."[11, 12]

PATHOLOGIC FINDINGS. The capsule is ruptured, causing a large opening at the level of the tibial margin; the head of the fibula is avulsed and displaced upward with the biceps attachment; the iliotibial tract may be ruptured; and often the cruciate ligaments are damaged. The peroneal nerve is stretched or completely torn.

TREATMENT. The ligament is relatively unimportant for stability of the knee. However, repair of the nerve is urgent. Early suture offers the best chance for recovery, whereas delay almost invariably results in failure. The capsule and the fibular head are repaired at the same time.

RUPTURES OF THE QUADRICEPS APPARATUS

Interruption in the continuity of the extensor apparatus of the knee takes place in the muscle, the quadriceps tendon, *the tendo-osseous junctions* at the upper and the lower poles of the patella, the patellar tendon and at the point of insertion of the patellar tendon into the tibial tubercle. The mechanism of injury includes: (1) *direct trauma*, a violent impact upon the affected structure, or (2)

[11] Platt, H The ligamentous-peroneal nerve syndrome, J. Bone & Joint Surg 10 403, 1928, Lancet 2.612, 1940.

[12] Highet, W. B, and Holmes, W Traction injuries to the lateral popliteal nerve and traction injuries to peripheral nerves after suture, Brit. J. Surg. 30:212, 1943

indirect trauma, usually an intense contraction of the quadriceps while the knee is forcefully flexed, e.g., stumbling while descending stairs. In the healthy, vigorous, athletic young adult, the tendon is strong, and rupture occurs within the muscle fibers or at the tendo-osseous junction, the latter often associated with avulsion of bone. With advancing age, tendinous structures become degenerate and fragile; therefore, in the aged individual, disruption of the tendon is more likely than muscle tears or rupture at the tendo-osseous junction. Fracture of the patella is not within the province of this text.

Fresh Rupture of the Quadriceps Tendon

Aged individuals are affected. Rupture is caused by forcible flexion of the knee while the quadriceps is strongly contracted. The patient often gives a history of stumbling while descending stairs, experiencing severe pain, buckling of the knee and falling down. Examination reveals moderate swelling, tenderness and ecchymosis immediately above the patella. A transverse sulcus is palpable instead of the prominent, rigid, quadriceps tendon. The patient is unable to extend the knee actively. If the rupture is limited to the tendons of the rectus femoris and the vastus intermedius, weak extension is effected by the vasti medialis and lateralis. The knee joint itself is not swollen, because blood and synovial fluid escape from the suprapatellar pouch.

The tear is transverse and involves the entire thickness so that the suprapatellar pouch is exposed. It extends medially and laterally into the aponeuroses of the vastus medialis and lateralis The margins of the tear are ragged and soft.

Treatment. The defect must be repaired immediately. Otherwise, the muscle retracts, and primary repair is rendered difficult. Mattress sutures of braided silk, immobilization for 6 weeks, and postoperative quadriceps and flexor exercises provide a very satisfactory result.

If rupture takes place close to the upper pole of the patella, the sutures are drawn through 2 drill holes made in a longitudinal direction in the patella.

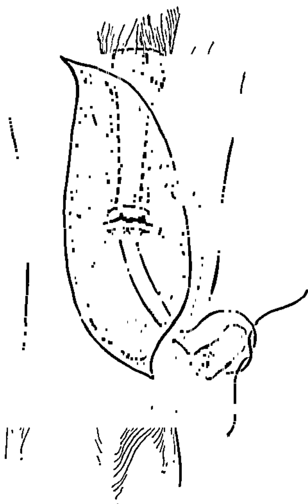


Fig. 419. Repair of old rupture of the quadriceps tendon. In addition to mattress sutures approximating the tendon ends, a tension suture of heavy braided silk is necessary to counteract the strong muscle pull. The tension suture is extracted after 6 weeks by means of a pull-out wire placed proximally.

Old Rupture of the Quadriceps Tendon

If the rupture has been neglected, the muscle contracts, and wide diastasis exists between the tendon ends. Inadequate bridging of the gap by proliferation of the tendon end takes place. In effect, the tendon is lengthened, and muscle contraction is rendered ineffective. A gap is palpable and, when the patient contracts the quadriceps, is often visible.

Treatment. Because of the vast expanse of muscle, it is possible to exert traction distally and overcome the separation between the tendon ends. An incision is made longitudinally through skin and the fascia over the anterior aspect of the lower half of the thigh, then

skirts the medial border of the patella. The scar tissue is excised, and tendon ends are freshened. Beginning at the middle third of the thigh, a long suture of stainless steel wire is inserted near the outer border of the rectus femoris and is extended distally by multiple insertions deep in the muscle belly until the strand emerges from the end of the proximal tendon fragment. Then it enters the distal tendon, emerges at the upper border of the patella, runs anterior to the patella and goes out through the skin over the anteromedial aspect of the tibia. The other strand of wire is inserted transversely through the muscle belly at the level of origin of the first strand and emerges at the medial border of the muscle Then, similar to the first strand, it is passed distally and emerges through the skin ½ inch alongside the other wire. A pull-out wire is attached to the proximal loop of suture and brought out through the skin of the thigh about 4 inches above the upper end of the incision. While an assistant, using towel clips, pulls the proximal tendon distally, the 2 strands are pulled up taut and tied over a rolled-up dressing or a large button. The tendon ends are approximated with braided silk mattress sutures Postoperatively, a plaster cast is applied with the knee in full extension. Four weeks later, gentle assistive, followed by active, flexion and extension exercises are started. At 6 weeks, the wire suture is removed

Avulsion of Patellar Tendon from Tibial Tubercle

Young, athletic individuals are affected. The patient stumbles, experiences a severely painful snap and falls to the ground Loss of active extension is immediate. The infrapatellar area is swollen, but on deep palpation a definite gap is perceived where normally lies the tense, resistant patellar tendon. Tenderness at the tibial tubercle is extreme. The patella lies at a high level If the tubercle is avulsed, x-ray films reveal the upward displaced fragment, the defect in the tibia and a high-riding patella.

Treatment. The tear usually extends medially and laterally through the patellar retinaculum. The patellar tendon is pulled down and is fixed by a braided silk suture through a drill hole made transversely in the anterior aspect of the tibia. The bone must be curetted before the tendon is approximated. If a large fragment has been avulsed, it is fixed with a screw. If small, it is removed. The patellar retinaculum should be left unsutured because suturing might limit flexion of the knee. Postoperatively, the knee is immobilized in extension for 6 weeks, followed by exercises.

Avulsion of the Patellar Tendon from the Patella

This injury affects the young patient, usually while participating in athletics. The tendon is torn from the lower pole of the patella, often avulsing a small segment of bone. The tear extends into the medial and lateral patellar retinaculum. Pain and loss of active extension are immediate. There are swelling and tenderness beneath the lower pole of the patella where a gap is palpable. Roentgenograms reveal a high-riding patella and perhaps a small fragment of bone.

Treatment. The avulsed fragment is excised. The lower pole of the patella is freshened. Two drill holes are made longitudinally in the patella. A crisscross suture of braided silk is passed through the tendon, and the strands are threaded through the holes and pulled up tight and tied while the patella is pulled distally. The patellar retinaculum is left unsutured. Postoperatively, the knee is immobilized in extension followed by exercises.

Old Ruptures of the Patellar Tendon

Shortening of the quadriceps makes approximation difficult This situation should be handled by the procedure described above for old ruptures of the quadriceps tendon.

Rupture of the Quadriceps Muscle

The fibers of the quadriceps may be torn by a violent contraction of the muscle or, more commonly, by severe direct trauma while the muscle is contracting. The young individual is injured, often while participating in an athletic game. He experiences excruciating pain, particularly if he attempts to extend the knee or when flexion is forced. Within a few hours, the thigh becomes markedly swollen and tense The swelling is very tender.

At this stage, a hemorrhage has occurred, and the muscle fibers are frayed. Repair at this time is impossible and likely to fail.

Conservative treatment is advisable. The patient is placed at bed rest, the knee is immobilized in full extension in a blanket splint, and icebags are applied in an attempt to reduce the bleeding. In a few days, heat is applied to promote absorption of the hematoma. Gentle knee exercises are started to prevent fixation of the muscles by cicatrix. Surgical intervention and forceful manipulations are contraindicated in the early stages, because the development of myositis ossificans would be encouraged.

The muscle tear heals by scar tissue. If the injury is not extensive, cicatrix will be minimal, and normal muscle function will be restored. When damage is extreme, the muscle belly separates, huge cicatrix forms; particularly if the rectus femoris is involved, fixation of the muscle to the sheath will restrict gliding. The range of flexion will be restricted, and extension will be weak A gap in the muscle may be palpated. It becomes necessary to remove the scar tissue and approximate the muscle by strips of fascia lata or by adapting the technic described for old ruptures of the quadriceps tendon.

Injury to the quadriceps, particularly the vastus medialis, often results in the development of myositis ossificans. This appears in roentgenograms as a diffuse, ill-defined, radiopaque shadow occupying the muscle shadow. Gradually, the shadow increases in size, then recedes, becomes increasingly dense and well outlined, and finally reaches the stationary, quiescent stage. Removal of the bony mass is foolhardy until the bony structure is well localized and no further activity is ascertained.

DISLOCATION OF THE PATELLA

The posterior articulating surface of the patella is smooth, covered with cartilage, oval in shape and divided into 2 facets by a vertical ridge which fits into the groove on the femur between the 2 condyles. When the patella is poorly developed and the vertical ridge is shallow, displacement out of the femoral groove takes place with ease. Repeated injury to the under surface occurs with dislocations and even more frequently with subluxations and results in erosion of the cartilage, friction of the bone, sclerosis and eventually a degenerative arthritis of the patellofemoral articulation which spreads to the rest of the knee joint. Recognition of this condition early and the removal of the patella rather than reconstructive procedures will prevent an irremediable disabling arthritis. If the subpatellar degeneration is not severe, resection of the abnormal tissue combined with the following procedures is acceptable.

Congenital Dislocation

Persistent dislocation of the patella at birth is frequently undiagnosed until it is apparent that the infant keeps the knee in flexion and is averse to moving the extremity. Roentgenograms are of no value inasmuch as ossification does not take place for at least 2 years and sometimes as late as 6. However, usually there is an associated knock-knee deformity, and the leg is rotated externally in relation to the thigh. The patella is palpable lateral to the lateral condyle of the femur and it cannot be replaced manually back to the femoral groove. The first phase of treatment is correction of the valgus deformity. In infancy, this can be handled satisfactorily by successive manipulations and casts. Later, however, osteotomy in the supracondylar area and internal rotation of the tibia are necessary.

At operation, the patella is found to be small and apposed and anchored to the lateral aspect of the lateral femoral condyle by the shortened fibers of the vastus lateralis component of the quadriceps aponeurosis. The main central quadriceps tendon is thick and well developed, and the vastus internus component is thinned and stretched. The anterior aspect of the lateral femoral condyle is shallow. The tibial tubercle insertion of the patellar tendon is found lateral to its usual site because of the tibial external rotation. The operative procedure consists of severing the lateral attachment of the vastus and displacing the patella medially. The defect left in the lateral quadriceps expansion is filled in by tendinous tissue removed from the elongated medial expansion. If this is insufficient, the semitendinosus tendon may be attached to the patella.

Recurrent Dislocation

The factors responsible for repeated dislocation are trauma and some pre-existent abnormality which permits lateral displacement. The trauma is usually an outward torsion of the leg while the knee is fully ex-

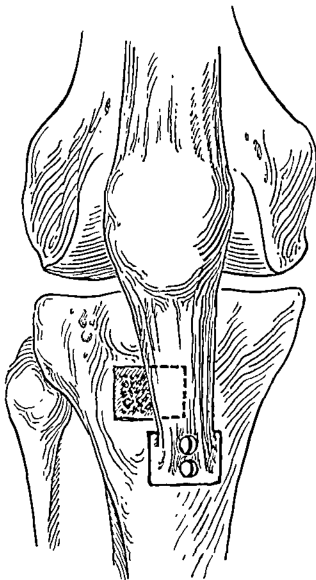


FIG. 420. Transplantation of the tibial tubercle for recurrent dislocation of the patella



FIG. 421. Recurrent dislocation of the patella. Lateral x-ray view shows a high-riding patella, evidence of abnormal elongation and laxity of the patellar tendon. This is one definite factor in dislocation and must be corrected at surgery.

tended, carrying the patellar tendon laterally. An example is the baseball player in the movement of swinging a bat. The first dislocation typically occurs in an adolescent; it is acute, is accompanied by severe pain, and the individual falls to the ground. The deformity is obvious. The bony prominence of the patella is over the lateral aspect of the knee, and the knee is held in slight flexion. Reduction is effected by extending the knee and flexing the hip, thereby relaxing the quadriceps muscle. Then the patella is reduced over the lateral condyle. Pain is relieved immediately. The dislocation must of necessity be accompanied by tearing away of the capsular structures which attach to the medial border of the patella. As a result, a moderate amount of soreness and tenderness is experienced at this site. Bleeding into the joint cavity causes distention of the knee with bloody synovial fluid. Subsequently, the intra-articular effusion subsides, and the medial aponeurotic fibers reattach themselves to the patella by weak and attenuated tissue which permits recurrences of lateral displacement. Frequently, the patient complains of slipping of the knee cap and catching over the lateral femoral condyle as though the patella were about to dislocate completely. Eventually, in many cases the dislocation recurs, and the same clinical picture is produced. With an increasing number of dislocations, the apposing aspects of the patella and the lateral femoral condyle become eroded and thinned, and the medial aponeurotic structures are greatly stretched. The patellar tendon may become elongated. Redisplacements occur with greater ease and less pain and effusion. For

the same reasons, reduction becomes easier, and the patient may learn to manipulate the patella back into place. Surgical correction is necessary to eliminate disability.

The theoretical and actual factors which contribute to patellar dislocation are: (1) genu valgum, (2) shallow lateral femoral condyle, (3) elongated patellar tendon evidenced by a high riding patella, (4) deficient vastus medialis, (5) external tibial torsion and/or lateral insertion of the patellar tendon, (6) deformed patella, and (7) shallow patellar groove on the femur.

Treatment

Surgical treatment is directed at establishing a straight quadriceps mechanism which keeps the patella medially and at eliminating all predisposing factors. Osteotomy of the femur is necessary for severe valgus deformity.

Hauser Operation. The tibial tubercle and its attached patellar tendon are transplanted medially and distally. This is the most effective procedure for severe repeated dislocations.

Campbell Operation. A pedicled flap is elevated from the medial capsule, threaded through the quadriceps tendon and attached to the adductor tubercle, thereby creating a check ligament. The defect in the capsule is imbricated.

Goldthwaite Operation. The patellar tendon is split longitudinally, and the lateral half is drawn medially where it is attached to the medial aspect of the tibia.

Tendon Transplantation. One of the inner hamstrings may be attached to the inner border of the patella.

Albee's Operation. This is indicated when the lateral femoral condyle is shallow. An osteotomy of the lateral condyle is done in a frontal plane, and the anterior fragment of bone is elevated and maintained in this position by a supporting piece of bone inserted beneath.

CHONDROMALACIA PATELLAE

The articular cartilage of the patella is relatively thick (7 mm.) as compared with femoral cartilage (3 mm.). It is the site of the earliest and most profound changes in degenerative arthritis of the knee. Every knee joint shows progressive degenerative changes after the age of 15, and the soft cartilage is worn down, especially where friction and pressure is greatest, namely, in the patellofemoral interval. The superficial cartilage layer of the patella shows the earliest change.[13] Chondromalacia is the term applied to rapid erosion and fragmentation of cartilage in the young adult.

Pathology

Grossly, the cartilage loses its bluish-white shiny translucency and becomes yellowish, opaque and soft. Cracks and fissures appear, giving a frayed appearance to the surface. The cartilage becomes ground down, exposing the underlying bone which becomes sclerotic. Gradually, marginal osteophytes develop.

Microscopically, the cartilage loses its staining qualities and contains relatively few cells. The remaining groups degenerate, swell and are heaped together in small groups, clusters of cells, called the lacunae of Weichselbaum. The thickness of the cartilage lessens, and collagenous fibrils appear between the cell clusters. The cement zone loses its deep-blue color and evenness and becomes wavy and irregular. This is the area of reactivated endochondral ossification which can be seen best at the margins where osteophytes develop.

Clinical Picture

The patient complains of intermittent pain over the anterior aspect of the knee and momentary catching, especially when arising from a chair or when ascending or descending stairs. Occasionally, a sensation of stiffness is experienced, relieved by activity. Grating is palpable and audible over the patella with active movement, accentuated by pressure over the patella. Compression of the patella against the femur is painful. Passive motion occurs without grating and discomfort.

Roentgenologic Findings

Early, films are negative. An indentation of the cartilage may be revealed by air arthrography. Later, sclerosis, irregularity and spurring are demonstrable, and the patellofemoral interval is narrowed.

[13] Bennett, G A, Waine, H, and Bauer, W.: Changes in the Knee Joint at Various Ages, New York, Commonwealth Fund, 1942

Treatment

Conservative treatment is indicated in most cases. The knee is put at rest, if necessary by applying a cylinder cast with the joint fully extended. Occasionally, an intra-articular injection of Hydrocortone is beneficial. Later, an elastic bandage will restrict *excessive flexion. Kneeling, squatting and running* must be avoided. Stair-climbing is done one stair at a time with the knee kept straight at all times. Later, gentle graduated quadriceps exercises are prescribed.

Surgical treatment is demanded when symptoms are severe and disabling. The knee is explored through a parapatellar incision which is extended upward between the rectus and the vastus medialis The patella is turned over, and the cartilage is pared down to bone. Loose cartilaginous bodies must be searched for and removed The slightest flake will eventually become hypertrophied and cause locking and traumatic degenerative changes. If cartilage degeneration is more extensive than has been suspected, or marked eburnation and thickening of the patella are found, a patelloplasty or a complete patellectomy, as is described in the following section, is done.

HYPERTROPHIC PATELLA

Hypertrophic degenerative changes in the patella usually follow chondromalacia, particularly after trauma, with or without fracture, or they occur as part of the gradually aging process plus wear and tear. The process within the knee begins earliest and most intensely in the patella and spreads to the condyles, the menisci and the synovium.

Clinical Picture

The course is one of recurrences and remissions of pain, swelling, and discomfort on flexion and extension, usually precipitated by excessive standing and walking or direct trauma. Each episode lasts from weeks to months. Climbing and descending stairs is difficult. Examination reveals thickening of the deeper soft tissues, broadening of the patella, and hypertrophic bone at the margins. Motion is greatly restricted and is accompanied by marked grating. Pain is relieved by rest and heat and is accentuated by activity. Marked stiffness is experienced after rest and subsides after walking.

Roentgenologic Findings

The patella is thickened and sclerotic and spurred, these changes existing throughout the joint. Loose osseous bodies are seen, but many *nonvisualized cartilaginous joint mice* are also present. The patellofemoral interval is narrowed, and the opposing surfaces are dense and irregular.

Treatment

Mild cases are controlled by *conservatism*, including heat, rest, external support, salicylates, intra-articular Hydrocortone injections, and gentle graduated quadriceps exercises. Diathermy is destructive and ill-advised.

The surgical treatment varies, depending upon individual preference. If one believes that the patella forms a necessary fulcrum for strong quadriceps action, a portion of the bone must be preserved. The degenerated posterior portion of the patella is excised down to cancellous bone, which is covered with a flap turned up from the fat pad.[14] Or the patella may be completely excised.[15] Knee joint function is very satisfactory after removal. Extension may be complete and strong, or only a few degrees of extension is lost. Critics of this procedure state that even this minimal instability subjects the joint to continued strains and further degenerative changes.

Technic of Patelloplasty. A long parapatellar incision extends upward between the junction of the rectus and the vastus medialis and downward to the tibial tubercle. Loose joint mice are removed. Both menisci are removed, since they usually show degenerative changes The patella is turned over to visualize the articular aspect. The soft tissues are separated slightly from the edges by a scalpel, the continuity of the extensor tendon being maintained anteriorly. The patella is cut on the flat with a hand saw to one fourth its original

[14] Cave, E F., and Rowe, C. R.: The patella in derangement of the knee, J. Bone & Joint Surg 32A 542, 1950

[15] Berkheiser, E. J · Excision of the patella in arthritis of the knee joint, J.A M A 113·2303, 1939

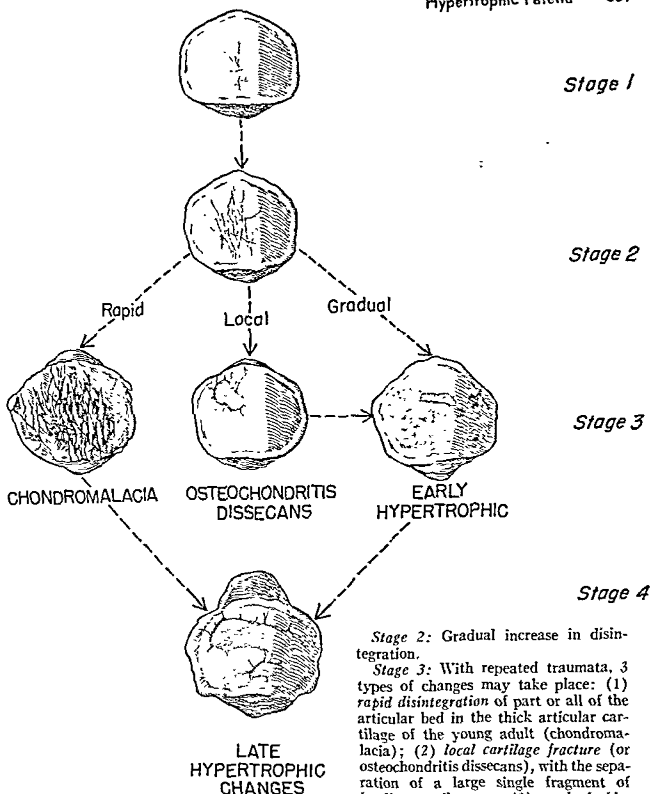


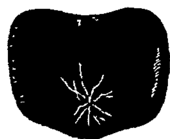
FIG. 422. Patellar changes.

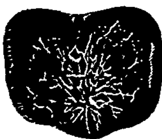
Stage 1: Represents the earliest articular change, usually a local softening of the cartilage, gradual unmasking of the vertical fibrils, and loss of hyaline matrix. Early fissuring or splitting of the cartilage may be seen.

Stage 2: Gradual increase in disintegration.

Stage 3: With repeated traumata, 3 types of changes may take place: (1) *rapid disintegration* of part or all of the articular bed in the thick articular cartilage of the young adult (chondromalacia); (2) *local cartilage fracture* (or osteochondritis dissecans), with the separation of a large single fragment of hyaline cartilage; or (3) *gradual thinning* of the entire articular cartilage, and erosion down to bone.

Stage 4: The "hypertrophic" or final stage, in which a large part of the articular cartilage has been lost, and the patella is broadened, thickened and irregular. (Cave, E. F., and Rowe, C. R.: The Patella: Its importance in derangement of the knee, J. Bone & Joint Surg. *32A:542-554*)



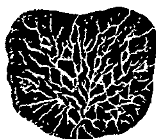




Fig. 423. Articular changes in chondromalacia of patella. (Cave, E. F., and Rowe, C. R : The patella: its importance in derangement of the knee, J. Bone & Joint Surg. 32A:542-554)

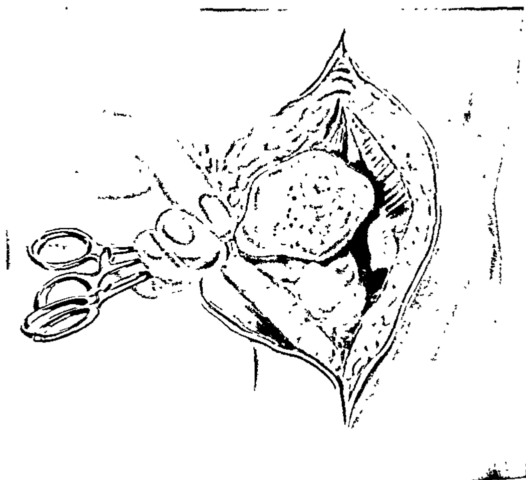


Figure 424 A (*Caption on facing page*)



FIG 424 (*Continued*). (A, *on facing page*) The patella is turned upward and is supported in this position. This gives adequate exposure of the patella and the infrapatellar fat pad (B, *above*) Showing partial resection of the patella. The joint is walled off with moist saline sponges during this process. Inset shows approximate line of resection. The major portions of the quadriceps and the patellar tendons are left intact.

(*Continued on p. 672*)

size. A flap of infrapatellar fat pad and synovial membrane is turned up from below to cover the raw bone and is sutured to the soft tissues at the patellar margins. This allows early smooth motion of the patella. Postoperatively, a plaster mold is applied, and quadriceps exercises are started on the second day. Resistive exercises and walking with crutches are begun on the 7th day. Exercises and improvement continue over a period of one year.

LARSEN-JOHANSSON DISEASE OF THE PATELLA

(Osteochondritis of the Poles of the Patella)

This syndrome occurs in adolescents, usually boys, between the ages of 10 and 14. Pain, soft tissue swelling and tenderness are noted over the lower, less commonly the upper, pole of the patella. It causes a limp and inability to kneel and run. Extension of

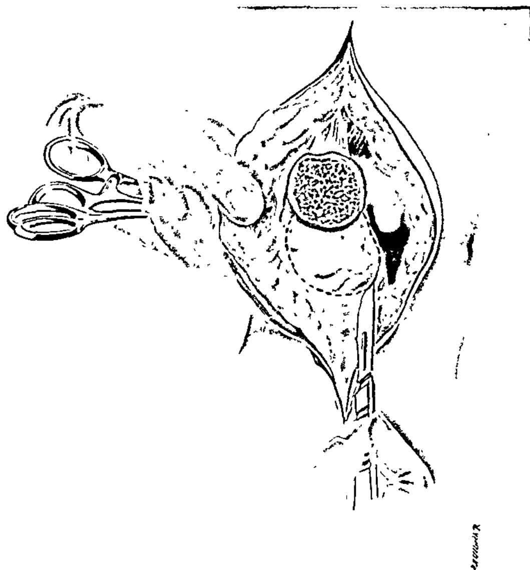


FIG. 424. (C) Approximately one quarter of the patella remains. The bone surface is covered by an infrapatellar flap of synovial membrane and fat pad. It is outlined by sharp dissection and is undercut back to the lower border of the patella. *(Continued on facing page)*

the knee against resistance accentuates the pain.[16, 17]

[16] Sinding-Larsen, M. F.: Acta radiol. 1:171, 1921-1922.

[17] Johansson, S · Ztschr. Orth. Chir. 43:82, 1924.

ROENTGENOLOGIC FINDINGS

Early, nothing abnormal is noted. Later, an area of irregular ossification develops over the affected pole of the patella. This is displayed

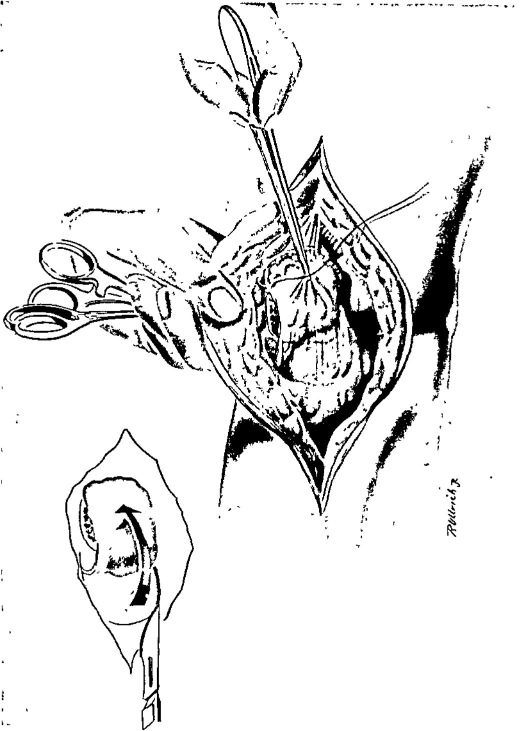


FIG. 424. (D) The patellar surface is covered by turning the infrapatellar flap upward and sewing it with fine cotton or silk sutures to the synovial membrane and tendinous cuff of the lateral, the medial and the upper borders of the patella (Cave, E. F., and Rowe, C. R.: The patella; its importance in derangement of the knee, J. Bone & Joint Surg. 32A:542-554)

as blurring, irregularity and small islands of condensation in a matrix of rarefied bone.

The cause is unknown. It is possible that repeated strains at the attachment of the patellar tendon or quadriceps tendon interfere with normal ossification, perhaps by interference with the blood supply. The condition is often associated with Osgood-Schlatter disease.

Treatment

Healing is spontaneous within a matter of a few weeks. A cylinder cast applied with the knee in extension relieves symptoms and may expedite recovery.

TRAUMATIC PREPATELLAR NEURALGIA[18]

Following direct trauma to the front of the knee, a persistent toothachelike type of pain or an electric-shock discomfort becomes a source of constant annoyance to the patient. Slight pressure, even the contact of clothing, accentuates the pain. The pathology is a contusion of the neurovascular bundle which issues from the skin in the prepatellar area and enters the prepatellar bursa. The nerve becomes fibrosed and adherent to surrounding structures.

Clinical Picture

Trauma to the front of the knee is followed by a transient prepatellar swelling. Within a few weeks, the neuralgic pain develops, and the slightest touch causes it to become excruciating. A well-localized point of tenderness is found over the middle of the outer border of the patella, the site of the neurovascular bundle. The pain persists, and the patient complains of inability to kneel, climb stairs, etc.

Treatment

Injection of a local anesthetic over the tender area often results in complete relief. Otherwise, it is necessary to remove the neurovascular bundle.

OSTEOCHONDRITIS DISSECANS

Osteochondritis dissecans, defined, is a condition of unknown etiology, occurring in children and young adults, and characterized by the separation of a fragment of bone and overlying cartilage from the articular surface.

Etiology

The condition is idiopathic. The following theories have been advanced:

1. **Trauma.** In the classic site at the inferior aspect of the medial femoral condyle, an impact from the tibial spine theoretically can occur with violent internal rotation of the tibia. In osteochondritis dissecans, the tibial spine is invariably prominent and likely to impinge repeatedly against the medial femoral condyle. Smillie explored a knee joint shortly after injury and detected a hairline crack in the cartilage opposite the tibial spine. The underlying fragment of bone was completely separated. When the lesion is located on the inferior articular aspect, it is considered due to impingement against a torn cartilage, which is an invariably associated lesion.
2. **Circulatory obstruction** by a thrombus or an embolus. This is unlikely in view of the frequent bilateral incidence.
3. **Low-Grade Inflammation.** However, no microscopic evidence can be found.
4. **Endocrine imbalance**
5. **Hereditary factor**

Pathology

The picture is one of aseptic necrosis. By some factor, probably trauma, a small fragment of bone is deprived of its blood supply. The bony architecture is unaltered, but the osteocytes disintegrate, and marrow elements are transformed to an amorphous debris. The surrounding bone undergoes hyperemia and decalcification. Capillaries invade the dead bone at the periphery, bringing young fibrous tissue, phagocytes which absorb the debris, osteoclasts which resorb the dead bone, and osteoblasts which deposit osteoid seams on the disappearing necrotic bone. Eventually, the necrotic bone is completely replaced by creeping substitution. In the meantime, the overlying cartilage remains viable, receiving its nutrition from the synovial fluid

More often, constant joint motion and repeated impacts cause micromotion of the fragment which prevents invasion and reconstruction. The overlying cartilage degenerates and

[18] Gordon, G. C. Traumatic prepatellar neuralgia, J Bone & Joint Surg 34B:41, 1952

tears, and the fragment is extruded into the cavity. *The loose body may remain attached* to its original site by a pedicle of synovial tissue, it may lie free within the joint, or it may gain reattachment to the synovium at a distance from its origin. Enlargement of the body takes place by hypertrophy of the cartilage which continues to derive nourishment from synovial fluid.

The residual cavity from which the bone was extruded fills in with fibrous tissue which is converted to fibrocartilage. A permanent *surface irregularity is the rule, leading eventually* to a variable degree of traumatic degenerative arthritis.

The usual location is the lateral wall of the medial femoral condyle adjacent to the attachment of the posterior cruciate ligament. Occasionally, it may be found on the lateral femoral condyle and posterior aspect of the patella in the lower medial quadrant. Lesions on the inferior articular aspect of a condyle, where it is subjected to repeated trauma *against the anterior end of a longitudinal* meniscal tear, do not penetrate deeply, involving only the articular cartilage and only a thin flake of bone.



Fig. 425. The femoral intercondylar notch view. Positioning of extremity. The tube is angled slightly upward. The lesion in osteochondritis dissecans is most often visualized in this projection.

Clinical Picture

Age. Children and adolescents are affected.

Sex. Males are predisposed.

History of trauma. This is rarely elicited.

Location. The lateral aspect of the medial femoral condyle near the attachment of the *posterior cruciate ligament is by far the most* frequent site. Bilateral involvement is not unusual. The lesion may be situated in other areas of the knee, in the elbow and the ankle, although rarely any other joint may be affected.

Associated lesions. Other types of osteochondritis are often found, such as Osgood-schlatter disease, Larsen-Johannson disease of the patella, and osteochondritis of the vertebrae.

Symptoms

The onset is insidious. Vagueness of complaints is characteristic. Pain is aching, intermittent and poorly localized. It is persistent, even at rest, and aggravated by exercise. A sensation of stiffness is *often experienced.* Locking, when momentary, is caused by a loose body being trapped between the condyles; it is relieved by the patient's kicking or maneuvering the leg. Persistent locking is due to the frequently associated torn meniscus.

Findings

Atrophy of the quadriceps, although minimal, is a constant sign. Well-localized tenderness is found by deep pressure over the femoral condyle with the knee flexed just beyond a right angle. The loose body may be perceived by palpation.

Roentgenologic Findings

If the lesion is recent so that rarefaction of the surrounding bone has not yet developed, or if the fragment consists solely of cartilage, the roentgenogram is negative. Later, a crater of rarefaction affecting the articular subchondral bone is visible. The crater contains a body which appears dense *in contrast with* the surrounding rarefied bone. The most common location in the anteroposterior plane, seen in the lateral roentgenograms, is just posterior to the middle of the condyle. In the anteroposterior view, it is located, as a rule, in the lateral aspect of the medial femoral condyle, often extending to the margin of the

intercondylar fossa. A postero-anterior tunnel view reveals the lesion best in this location. This is done by placing the patient prone, flexing the knee to 60° and directing the tube in a caudad direction so that the rays intercept the joint at an angle of 30°. Another method is by placing the patient in the supine position, flexing the knee to 45° over a casette and directing the rays perpendicular to the film.

Defects involving only the articular cartilage can be visualized by air arthrography. When the loose body contains an osseous center, it is seen readily. Otherwise, contrast air arthrography is necessary. Several particles of cartilage extruded into the joint may attain large proportions, but only one crater is present.

Treatment

In the early stages, before extrusion of the fragment, resorption of the necrotic bone and replacement without involvement of the overlying cartilage is possible if the joint is kept at absolute rest without weight-bearing for several months to a year. This is especially true in children.[10] A cylinder plaster cast is applied, and crutch ambulation is permitted. If the condition is bilateral, absolute bed rest or confinement to a wheel chair is advisable Progress is followed by roentgenograms. Restoration of bony architecture and excellent joint function can be expected. If the condition progresses to extrusion of an osseocartilaginous body, the joint mouse must be removed and the cavity curetted before repeated incidents of locking irreparably damage the articular surfaces. Some orthopaedic surgeons prefer to excise the lesion even before extrusion, but often the overlying intact cartilage must be removed, and the inevitable result is an irregular joint surface with symptoms of degenerative arthritis. The older the patient and the longer the history of symptoms, the more likely is the cartilage to have been damaged and extrusion of a loose body probable. In such cases arthrotomy is advisable.

[10] Green, W T, and Banks, H. H.: Osteochondritis dissecans in children, J Bone & Joint Surg. 35A:26, 1953.

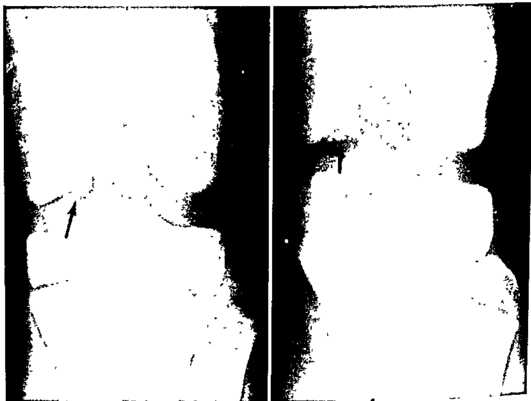


Fig. 426. Osteochondritis dissecans. The separating bone island was not visualized in the ordinary anteroposterior views. At surgery, the lesions were completely covered by articular cartilage, but their site was detected by surface irregularities.



Fig. 427. So-called "Osgood-Schlatter disease." The epiphysis extends downward anteriorly toward the site of the tibial tubercle and is attached to the shaft by a similar extension of the cartilaginous epiphyseal plate. The patellar tendon attaches to the tubercle and in turn to the shaft of the tibia by a weak link of cartilage. During the stage of active growth at puberty, the epiphyseal plate is thickened, but its junction with bone is rendered insecure by proliferation and maturation of chondrocytes. Strong quadriceps pull partially avulses the tubercle. The tonguelike extension of epiphysis frequently remains unossified or contains one or several small ossification centers and the traumatic nature of this condition is not recognized. The tibial tubercle may develop separately from the epiphysis, but the mechanism of production of symptoms is the same.

Technic. The joint is exposed through an anteromedial incision. This may be extended upward if thorough exposure is necessary. The loose body is removed. The cavity is gently curetted, and the overhanging edges of cartilage are cut away. The underlying meniscus is examined and, if torn, is removed.

The necrotic fragment may lie within its crater covered by normally appearing cartilage However, by probing the area about the site of attachment of the posterior cruciate, the cartilage over the necrotic site is soft and doughy as compared with the normal, rubbery, resistant cartilage. Or the overlying cartilage may be dull, yellow in color and fibrillated instead of shiny, bluish-white and smooth. To localize the lesion accurately at surgery, a needle is inserted into the suspected area, and a roentgenogram is made. Often the degenerate cartilage is elevated and surrounded by a depressed rim. Then an incision is made in the cartilage around the nonviable area, and the loose body is removed with its covering of cartilage. The cavity is curetted down to normal bleeding bone.

Postoperatively, weight-bearing is delayed for several weeks to several months, depending on whether or not the lesion corresponds to the weight-bearing surface of the femoral condyle. If it is over the most inferior aspect, complete filling in of the defect as noted in roentgenograms is mandatory before weight-bearing is permissible. Similarly, quadriceps exercises should be limited.

OSGOOD-SCHLATTER DISEASE (Osteochondritis of the Tibial Tubercle; Strain or Partial Avulsion of the Tibial Tubercle)

Defined, Osgood-Schlatter disease is a painful disabling swelling about the tibial tubercle, occurring in adolescents.[20, 21]

Etiology

Trauma is a frequent factor, particularly a single violent or lesser repeated flexion of the knee against a tight quadriceps. Children about the rapid growth period of puberty, particularly boys, are predisposed. The condition is often bilateral.

[20] Osgood, R. B.: Lesions of the tibial tubercle occurring during adolescence, Boston M. & S. J. 148·114, 1903

[21] Schlatter, C.. Verletzungen des Scheinbeinformigen Fortsatzes d Oberen Tibia Epiphyse, Beitr z. Klin Chir. 38·874, 1903.

Clinical Picture

Pain, tenderness and soft-tissue swelling without inflammatory signs are well localized to the tibial tubercle. Activity imposing strong quadriceps contraction and therefore strain upon the tubercle aggravates the discomfort, e.g., climbing stairs and running. Kneeling is painful. Active extension of the knee against resistance is painful. The course is chronic and recurs over a period of months to several years, but usually it ceases at or before 18 years of age, when the apophysis fuses to the main bone. Occasionally, the symptoms may persist into adult life.

Roentgenologic Findings[22]

The tibial tubercle consists of multiple fragmented appearing areas of ossification which are dense in contrast with the underlying osteoporotic area in the main bone. The ossification in the tubercle may be single and in continuity with the ossification center of the upper tibial epiphysis which is prolonged distally as a tongue-shaped process. The soft tissues anterior to the tubercle are swollen, particularly the shadow of the patellar tendon which may contain an irregular area of calcification or well-circumscribed bone. These findings are in contrast with actual complete avulsion of the tubercle in which the ossification center is displaced upward.

Pathology[23]

The tibial tubercle develops as an extension of the upper cartilaginous epiphysis, which is prolonged anteriorly and distally. A single, occasionally a double, ossification center develops within the tubercle and generally fuses to the center of the main epiphysis at the age of 16 It fuses to the main bone at 18. Before that time, its attachment to the parent bone is by a layer of proliferating cartilage which is an extension of the epiphyseal growth plate. Beneath the growing cartilage, the newly formed bone forms a soft weak link which yields to the pulling strain of the quadriceps Actual complete separation can occur. More frequently, however, the separation is minimal but sufficient to obliterate the blood supply to the tibial tubercle. The latter undergoes aseptic necrosis. The neighboring bone undergoes active hyperemia, which is manifest by osteoporosis. Capillaries and phagocytes invade and remove the dead bone, and the picture of fragmentation results. Eventually, the new bone is formed and fuses to the main bone. The description of pathology and pathogenesis can be applied to all of the osteochondritides of the epiphyses. They all occur during the rapid growth period.

Treatment

Conservative treatment is effective in most cases. This consists of maintaining the knee in full extension, preferably by a cylinder cast, for several months. This removes the pull of the quadriceps and permits revascularization and reossification of the tubercle

Surgical. If the course is prolonged and the disability persistent, surgical intervention is indicated. Drill holes made through the tubercle into the main bone form channels through which rapid revascularization can occur. Pain and disability are relieved permanently within a few weeks. Bone pegs removed from an adjacent area of the tibia may be inserted through the drill holes, providing an immobilizing bone graft.[24] If the tubercle forms an abnormally large prominence which is cosmetically disfiguring and may be a source of pressure pain in the future, the protruding fragments may be removed through a transverse incision. The attachment of the patellar tendon extends well beyond this point, both distally and laterally, and is unaffected.[25]

Complications

The most common complication is failure of union of the tubercle to the tibia. This is the cause of persistence of symptoms into adult life. The ununited fragment should be removed. A less common but important sequel is premature fusion of the anterior end of the upper tibial epiphysis. This leads to genu recurvatum When the disability is prolonged, the patella may become high-riding and its

[22] Ferguson, A B · Roentgen Diagnosis of the Extremities and Spine, New York, Hoeber, 1945

[23] Luck, J V: Bone and Joint Diseases, p 152, Springfield, Ill, Thomas, 1950.

[24] Bosworth, D M. Lesions of the tibial tubercle and their treatment, Am J Surg. 43 526, 1939.

[25] Thomson, J E M Operative treatment of osteochondritis of the tibial tubercle, J. Bone & Joint Surg. 38A 142, 1956.

inferior facet subject to constant trauma and osteoarthritis.

DEFORMITY ABOUT THE KNEE

Bony structures may become deformed about the knee and, as a result, the femur and/or the tibia deviates in a corresponding direction. In the coronal plane, the knee is angulated medially (valgus) or laterally (varus). In the sagittal plane, the knee is angulated posteriorly (recurvatum) or anteriorly (antevertum).

Causes of Bony Deformity at the Knee

1. **Asymmetric rate of growth of epiphyseal plate**

A. *Trauma* causing premature closure of a portion of the plate

B. *Infection* can temporarily retard activity or permanently destroy the plate. Rarely, it can accelerate the rate of growth.

C. *Static abnormality.* A deformed shaft of the femur or the tibia or muscle imbalance brings excessive pressure to bear upon one end of the plate, thereby retarding its activity. Conversely, the opposite side of the plate continues to function uninhibited.

D. *Metabolic and nutritional disturbance,* notably rickets

E. *Osteochondrosis of the upper tibial epiphyseal plate* (Blount's disease)

2. **Malunion of fracture at the metaphysis**
3. **Developmental disturbances**

A. *Osteogenesis imperfecta*

B. *Chondrodysplasia*

4. **Congenital bowing of the tibia and the femur**

Principles

Premature closure of a portion of the epiphyseal plate causes asymmetric longitudinal growth throughout the growth period. Therefore, correction before growth has been completed is followed invariably by recurrence; this necessitates repeated surgery. Temporary inhibition of longitudinal growth, e.g., rickets, should await removal of the offending factor before correction is attempted. Other ill-understood conditions, as osteogenesis imperfecta, become inactive after puberty, whereupon surgical intervention may be attempted. Congenital bowing of the tibia is associated with some deficiency in bone healing. Osteotomy is fraught with the danger of nonunion. An osteotomy should not be attempted before the age of 4, when the epiphyseal structure becomes fully developed.

Valgus deformity occurs most commonly in the femur and is associated with external rotation of the distal end and varus deformity of the tibia. Correction necessitates not only a femoral osteotomy but also internal rotation of the distal fragment and, later, osteotomy of the tibia. Varus deformity takes place most often in the tibia and is associated with internal torsion of the distal shaft. The distal fragment must be rotated externally at the same time as osteotomy.

Osteotomy is performed at the site of maximum deformity, or the apex. One must avoid trauma to the epiphyseal plate. Deformity is corrected by (1) an *open wedge osteotomy* on the concave side. This tends to lengthen the bone and subjects the soft tissue, notably the peroneal nerve, to traction injury. (2) *Removal of a wedge* of bone from the convex side tends to shorten the bone. When lengthening of the bone is desirable, it is necessary to protect the peroneal nerve by transplanting it anteriorly. In addition, the Achilles tendon may require lengthening. Other shortened soft-tissue structures tend to cause some recurrence, so that the deformity must always be slightly overcorrected.

Temporary Epiphyseal Arrest[26, 27]

Haas has demonstrated that epiphyseal growth is arrested by encircling the epiphyseal plate with a wire loop. Normal growth is resumed when the wire is removed. Metallic staples may be used instead of the wire. Complete cessation of growth is obtained when 3 staples are used. Growth pressure is tremendous and will bend or break the staples when a lesser number is used. By enclosing one side of an epiphyseal plate, the opposite side continues to grow, and deformity is corrected. The method can be used only when some growth activity occurs on the unstapled side. Insertion and removal of the staples must be timed so that correction is maintained at the

[26] Blount, W. P., and Clark, G. R.: Control of bone growth by epiphyseal stapling, J. Bone & Joint Surg. 32A:464, 1949.

[27] Haas, S. L.: Retardation of bone growth by a wire loop, J. Bone & Joint Surg. 27:25, 1945.



FIG. 428. Genu valgum corrected by delayed osteoclasis. (*Left*) Original deformity. (*Center*) Partial osteotomy including all of the bone but the lateral cortex. The bone grafts in the wedged out area can be seen. (*Right*) After completion of correction by manual osteoclasis. The straight weight-bearing line has been restored.

conclusion of treatment. If the deformity is progressive, sufficient residual growth must remain to obtain correction. If deformity is stationary, stapling can be done anytime during the growth period and removal effected when correction is achieved. Premature closure of the epiphysis is a complication.

By stapling both sides of the epiphyseal plate, growth in length is restricted, and limb length may be balanced

Technic. The site of maximum deformity determines the epiphyseal plate to be restricted. A 1-inch longitudinal mid-lateral or mid-medial incision is made over the epiphyseal disk. The periosteum is elevated, and the edge of the epiphyseal plate is disclosed as a transverse white band which encircles the bone The distal femoral epiphyseal plate is saucer-shaped, with its dome extending distally. Therefore, each staple must be inserted so that two thirds of the crosspiece of the staple lies distal to the white line The proximal tibial epiphyseal plate is almost V-shaped, its dome extending proximally. Therefore, the staple must be placed in a more proximal position to avoid injuring the plate. One staple is placed centrally and the other two on either side. Stapling must not be done too far anteriorly or posteriorly, because a secondary deformity will be produced. A roentgenogram checks the position of the staples before closure of the wound. Postoperatively, a plaster cylinder may be employed for 3 weeks to encourage subsidence of surgical reaction. The child is permitted to walk immediately, unaided.

GENU VALGUM
(Knock Knees)

The deformity consists of medial angulation of the knee and outward deviation of the longitudinal axis of both the tibia and the femur. In advanced cases, the distal end of the femur and the tibia is rotated externally by the pull of the biceps femoris and the tensor fascia femoris, and the distal shaft of the tibia develops a compensatory internal

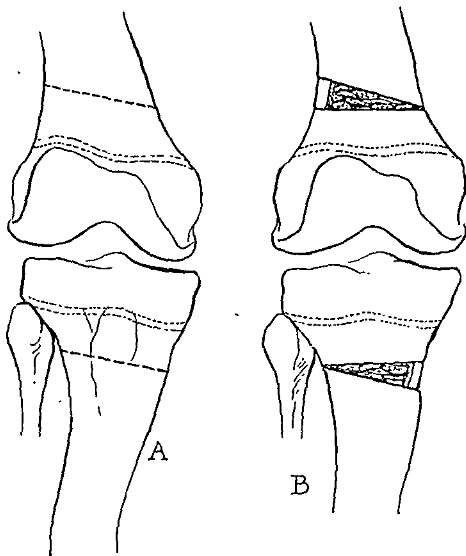


Fig. 429. Genu valgum, surgical correction. (*Left*) The valgus deformity usually takes place in the femur. A compensatory varus and internal torsion develops in the tibia. Both deformities must be overcome. (*Right*) Line of transection of bone is placed parallel to the epiphyseal plate. The open-wedge type of osteotomy shown here is preferred when gain in length is desirable.

torsion A commonly associated abnormality is a pronated and flat foot The most common cause is a fracture dislocation of the epiphysis or a fracture through the outer portion of the epiphyseal plate Rickets is now a rare cause, although a nutritional disorder must be considered, particularly when the deformity is bilateral. With subsequent growth, the medial condyle assumes larger proportions than the lateral condyle, and the articular surfaces lie at an oblique angle. The quadriceps extensor mechanism crosses the joint over the lateral aspect where the patella tends to subluxate or dislocate outward. On the lateral side, the soft-tissue structures, including the biceps, the iliotibial band and the peroneal nerve are shortened. On the medial side, the soft-tissue structures are elongated. The medial collateral ligament is stretched and lax, causing instability of the joint.

Treatment

Conservative. Mild cases in actively growing children may often be improved by conservative treatment. Pronation of the feet is corrected by elevating the inner border of the shoes from $\frac{3}{16}$ inch to $\frac{1}{4}$ inch. Knock-knee braces are worn continuously. These consist, in principle, of a lateral bar to which a leather knee cuff can be attached and tightened.

Surgical. More severe cases require surgical intervention. If the lateral portion of the epiphyseal plate is intact as seen in roentgenograms, it is assumed to contribute to longitudinal growth but at a reduced rate. This situation is suitable for stapling the medial portion of the epiphyseal plate. If correction is achieved before growth is complete, some overcorrection must be attained. After epiphyseal fusion, an osteotomy must be per-

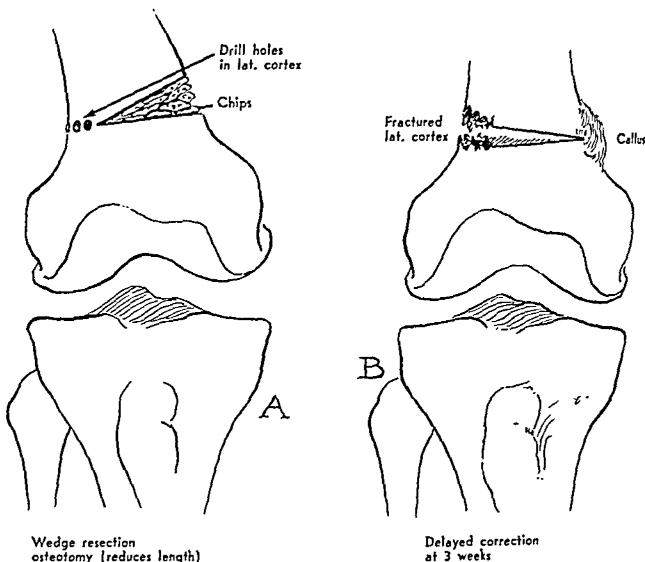


FIG. 430. Surgical correction of genu valgum by wedge resection osteotomy.

formed at the site of maximum deformity, which may be either in the femur or the tibia. If the length of the extremity is adequate, a wedge of bone is removed from the medial side. This produces some shortening. When the limb is already short, osteotomizing the outer side creates a wedge-shaped defect and lengthens the extremity. It may be necessary to lengthen the soft tissues on the lateral side and transplant the peroneal nerve anteriorly.

Technic of Open Wedge Osteotomy. A longitudinal incision is made over the outer aspect of the thigh just above the lateral femoral condyle. The bone is approached between the vastus lateralis and the lateral intermuscular septum and is exposed subperiosteally. The osteotome is driven through the bone, leaving the medial cortex intact. By displacing the leg medially, the defect is opened widely as the medial cortex is fractured. A piece of iliac bone is wedged into the defect to maintain the correction.

When valgus deformity is extreme and the limb is shortened, the technic must be varied. The osteotome must be aligned at an acute angle but parallel with the distal articular surface. Opening the wide defect during correction is resisted by the contracted soft tissues on the lateral side. Therefore, release of contracture must precede the osteotomy. The laterally displaced patella is mobilized by freeing the lateral quadriceps expansion from the lateral intermuscular septum. The iliotibial band is cut transversely, and a segment of the lateral intermuscular septum is removed above the epicondyle. The biceps tendon is lengthened by an oblique tenotomy. The lateral head of the gastrocnemius is dissected free from the posterior aspect of the lateral epicondyle. To avoid severe traction injury to

the peroneal nerve, it is transplanted anteriorly. The fibular origin of the peroneus longus is removed as the nerve is traced downward where it hooks around the neck of the fibula. This allows the nerve to be placed anteriorly. The oblique osteotomy is performed, and the lateral condyle is levered downward, opening an enormous wedge. External rotation of the condyles must be corrected at this time. A large piece of cortical bone is fitted into the defect and held with a screw.

Correction of a valgus and external rotation in the femur results in a converse varus and internal rotation of the tibia. An open wedge osteotomy is performed in the upper end of the medial aspect of the tibia, externally rotating the distal fragment at the same time. Additional lengthening is thereby secured. A piece of cortical bone is forced into the defect and held with a screw. A plaster cast is applied and held until union is complete.

Technic for Wedge Resection Osteotomy.[28] Through a longitudinal incision above the medial condyle, the femur is approached between the vastus medialis and the medial intermuscular septum. A cuneiform wedge of bone is removed, leaving the lateral cortex intact. Several drill holes are made in the lateral cortex. The cuneiform wedge of bone is broken up into small pieces which are packed into the defect. The periosteum is carefully sutured to hold the chips in place. Postoperatively, a plaster cast is applied About 3 weeks later, soft callus will have formed. A cylindrical section is removed from the cast and, under anesthesia, manual osteoclasis is performed. It may be possible to correct rotation at the same time. The defect is filled with plaster, and immobilization is continued until union is complete, usually 6 weeks in children and 10 to 12 weeks for adults.

This method is especially suitable for cases where possible nonunion is a consideration and when lower limb lengths are equal.

GENU VARUM
(Bowlegs)

This deformity consists of outward angulation of the knee joint, the longitudinal axis of the femur and the tibia deviating medially. The deformity involves the tibia alone or the femur, the tibia and the fibula. An internal torsion of the distal shaft of the tibia is an associated abnormality. This causes an intoeing of both feet. As the feet are pointed forward in walking, the patellae face outward, and the bowleg deformity appears to be accentuated.

Rickets, formerly the most common cause, is now almost extinct with the advent of vitamin D treatment. Some cases are congenital. Many are postural, caused by the infant's lying in the prone position with the thighs abducted and the toes turned inward. Injury to the medial portion of the upper tibial epiphyseal plate causes unilateral deformity. Rarely, developmental and endocrine disturbances are causative.

Tibia Vara[29]
(Osteochondrosis Deformans Tibiae; Blount's Disease)

This is a fairly common cause of genu varum. The medial portion of the upper tibial



Fig. 431. Genu varum. Etiology was vitamin-resistant rickets. Spontaneously corrected after administration of vitamin D in high dosage.

[28] Moore, J. R.: Osteotomy—osteoclasis. A method for correcting long bone deformities, J. Bone & Joint Surg. 29:119, 1947.

[29] Blount, W. P.: Tibia vara, J. Bone & Joint Surg. 19 1, 1937.

epiphyseal plate displays abnormal growth which results in delayed ossification of the medial portion of the epiphysis. A beaklike projection of bone develops medially and upward from the metaphysis. Irregular areas of cartilage situated within this metaphyseal outgrowth appear in roentgenograms as rarefaction. The outgrowth is capped with hyaline cartilage.

The *infantile type* occurs in an overweight child during the second year of life. The varus may become stationary or get progressively worse. X-ray films show (1) a sharp angular deformity immediately below the upper tibial epiphysis; (2) the medial side of the epiphyseal plate is widened and irregular; (3) the ossification center in the epiphysis is wedge-shaped, the base laterally and the narrow end medially; (4) the medial portion of the metaphysis displays a beaklike projection, containing multiple rarefied areas (cartilage); 50 per cent of cases are bilateral.

The *adolescent type* is unilateral and makes its appearance between 6 and 12 years of age. The x-ray findings suggest retarded activity of the medial portion of the upper tibial epiphyseal plate. The plate at this site is irregular, narrow, and sometimes obliterated. A sharp angular deformity develops in the metaphysis.

Clinically, bowlegs and a waddling gait are typical. The deformity below the knee is sharp. Constantly associated abnormalities include genu recurvatum, internal tibial torsion, and pronation of the foot. The medial tibial condyle displays a prominence. A slight amount of shortening is usually observed. Spontaneous correction may occur occasionally. The deformity does not increase after closure of the epiphyseal plate.

Treatment of Genu Varum

In the absence of a metabolic and endocrine factor or premature closure of the epiphyseal plate, spontaneous correction is possible. A bowleg brace is worn continuously. This consists of a leather cuff, attached to a medial bar, which encircles the knee and is tightened daily. Although no evidence of rickets can be found, vitamin D should be given. The author has observed rapid correction on this regimen. In some clinics, both limbs are immobilized in casts for a number of weeks to effect decalcification and, theoretically, to soften the bones prior to applying corrective braces.

Severe deformities require surgical correction. If asymmetric activity of the epiphyseal plate is continuing, treatment is delayed until growth has been completed. Roentgenograms are studied to determine the site of maximum deformity and the degree of correction. An open wedge osteotomy on the medial side at the point of maximum deformity is preferred, because it restores some length. In tibia vara the osteotomy passes transversely immediately below the epiphyseal plate. Danger of injury to the plate makes it advisable to wait until the epiphysis has closed. Then the osteotomy is done immediately below the epiphysis. The defect is widened medially and a little posteriorly and maintained by cortical grafts. Internal rotation of the tibia is corrected at the same time.

GENU RECURVATUM

Backward bowing of the knee joint is due most commonly to injury with resultant retarded activity of the anterior portion of the lower femoral or upper tibial epiphyseal plates. Consequently, the posterior portions of the condyles grow larger in contrast with the anterior portions, and the articular surfaces are correspondingly inclined in the forward direction. When the femur is involved, the articular surface and the epiphyseal line, as seen in roentgenograms, are inclined forward and upward in relation to the longitudinal axis of the shaft. In the case of the tibia, the articular surface and the epiphyseal line incline forward and downward. Regardless of the site of bony deformity, surgical correction is confined to the upper end of the tibia.

The deformity is also caused by muscle imbalance, as in poliomyelitis, and by equinus deformity of the foot which throws the knee into extreme hyperextension. This is discussed in the section on Poliomyelitis

Technic[30]

A transverse incision is made anteriorly just above the tibial tubercle. The upper end of the tibia is approached by a longitudinal inci-

[30] Brett, A. L. Operative correction of genu recurvatum, J. Bone & Joint Surg 17:984, 1935.

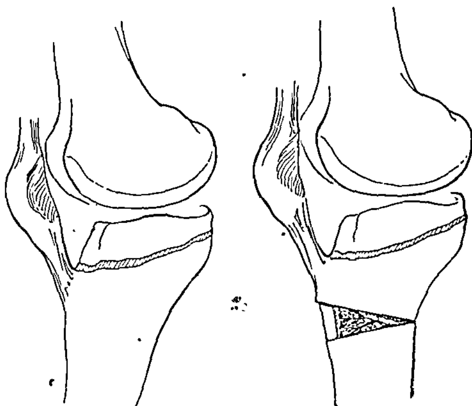


FIG. 432. Genu recurvatum, surgical correction. (*Left*) The deformity is usually caused by retarded longitudinal growth of the anterior portion of the epiphyseal plate, or by defective ossification of the posterior portion of the upper tibial epiphysis. The latter cause produces a wedge-shaped epiphysis. (*Right*) The line of transection of bone should parallel the epiphyseal line. An open wedge osteotomy, as shown here, is preferred when gain in length is desirable.

sion on each side of the patellar tendon. The bone is osteotomized transversely just below the capsular attachment. The proximal segment is wedged upward, and the defect is maintained by wedging strong cortical bone grafts into the gap anteriorly. Postoperative immobilization in a cast and abstinence from weight-bearing for about 12 weeks are required. Before this time, some recurrence is likely, due to strong quadriceps pull.

DEGENERATIVE JOINT DISEASE OF THE KNEE

The knee is the most commonly affected joint. The usual degeneration of advancing age is greatly accentuated under certain circumstances and extremely disabling. The condition is frequently preceded by:

1. **Injury** (torn menisci, fractures, dislocating or subluxating patella, joint mice). Irregular joint surfaces or loose bodies directly damage the cartilaginous surfaces.
2. **Strain** (obesity, valgus, varus). Abnormally increased pressure is brought to bear on the weight-bearing surfaces
3. **Infection** (suppurative, rheumatoid, tuberculous). Surface destruction results in mechanical incongruity.
4. **Central Nervous System Disease** (tabes)

Chondromalacia of the patella frequently initiates the condition. The patellofemoral joint becomes degenerated and eroded by friction. The irregular surfaces of the femoral condyles, in turn, damage the tibia.

CHARACTERISTIC FINDINGS

Painful creaking and grating on active motion, particularly about the patella, is an early finding even before roentgenograms become revealing. It is due to friction of the patellofemoral articulation. Passive motion relaxes the quadriceps, relieves patellofemoral compression and eliminates the subpatellar grating. The grating sensation and pain is accentuated by any activity requiring forceful contraction of the quadriceps, e.g., climbing stairs, descending stairs, arising from a sitting position, etc. Tenderness occurs about the joint margins where the swollen, congested synovium protrudes The finding should not be interpreted as due to a torn cartilage. If increased synovial fluid is present, the patella may be floating, and subpatellar grating and pain are absent. Muscle spasm accompanies attacks of acute synovitis. Muscle atrophy, particularly of the quadriceps, is a late finding when the destruction is advanced and motion is limited. In this late stage, the bony

margins are enlarged, the range of active and passive motion is greatly diminished, and a fixed deformity in partial flexion is usual. Attacks of acute inflammatory synovitis accompanied by increased effusion and muscle spasm are common. Locking may be caused by nipping of a hypertrophied synovial fringe, displacement of an easily torn meniscus, or jamming of a detached osteophyte.

Pathology

The basic gross and microscopic findings of degenerative arthritis have been described in the section on Arthritis. In addition, certain conditions characterize knee-joint involvement. The patellofemoral articulation is the site of maximum damage. An ulcerative deepening of the undersurface of the patella corresponds to a site of maximal erosion over a femoral condyle. Many flakes of cartilage detached from the joint surface are found. Also, a variable number of rounded cartilaginous or osteocartilaginous loose bodies may represent stages in the continued growth of the flakes; or they may be formed by metaplasia in the synovium and be detached later. Degeneration also affects the menisci, making them extremely vulnerable to injury. Minute to gross tears with displacement are not uncommon. Although participating in the joint deterioration, usually the cruciate ligaments do not succumb. They are generally found to be intact even in degeneration of extreme degree. The infrapatellar fat pad is thickened and somewhat indurated. It may hypertrophy and protrude medially where it may be mistaken for a tumor.

Treatment

Conservative. Severe degenerative changes as seen in roentgenograms are compatible with a painless, stable, functioning knee provided that the quadriceps is strong and the joint is protected from unnecessary strains and pressures. Severe pain and effusion denote temporary irritation which frequently can be controlled by the following measures:

1. *Avoid weight-bearing,* if necessary by crutches.
2. *Immobilize the knee* by a cast, a splint, a knee corset, a brace or an elastic bandage.
3. *Heat,* preferably moist heat. Short-wave diathermy is destructive and should be avoided.
4. *Hydrocortisone* is injected at weekly intervals.
5. *Quadriceps exercises* are performed daily to maintain strength of the main stabilizing factor of the knee.
6. *Procaine needling* of a localized tender spot over the joint interval frequently relieves pain quickly.

The failure to respond to conservative treatment is sufficient indication for surgical intervention.

Surgical Treatment. Locking, irritation, pain and effusion are caused by an osteophyte, a large flake of cartilage, a hypertrophied synovial fringe, an enlarged fat pad, or a torn degenerated displaced meniscus. Removal is indicated.

Patellectomy. Localized patellofemoral degenerative arthritis without involvement of the rest of the joint is an indication for removal of the patella. Failure to do this will eventuate in progression and damage to the entire knee. Tangential x-ray views of the patellofemoral articulation should be done routinely. The earliest signs of degenerative arthritis will be detected at this site, and patellectomy will prevent extension to the other joint surfaces.

Technic.[31] An incision is made skirting the medial or lateral border of the patella. The capsule about the patella, which is the continuation of the quadriceps tendon, is split longitudinally in the mid-line. The medial and the lateral sides of the patella are grasped with towel clips and displaced forward out of the wound. The patella is divided longitudinally through three fourths of its thickness by a hand saw, and section is completed manually or by a chisel. The two halves of the patella are spread, and the interior of the joint is inspected for loose bodies and other pathology. Next, the patellar halves are enucleated from the capsule by blunt dissection. The capsule is closed by imbrication. If the defect is weak, it can be reinforced by inserting strips of fascia lata or by sewing the quadriceps tendon to the patellar tendon by heavy gauge thread. Postoperatively, the knee is

[31] Boyd, H. B., and Hawkins, B. L.: Patellectomy, a simplified technique, Surg., Gynec. & Obst. 86:357, 1948.

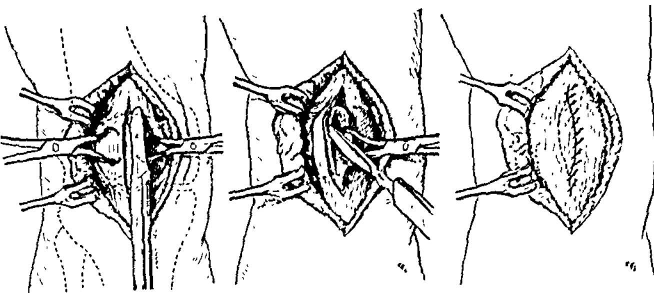


FIG. 433. Technic of patellectomy. (*Left*) Sawing patella in two halves. (*Center*) Enucleation from capsule. (*Right*) Imbrication of capsule. (Boyd, H. B., and Hawkins, B. L.: Surg., Gynec. & Obst. 86:357)

immobilized for 3 weeks, during which time quadriceps setting exercises are practiced. Thereafter, guarded active quadriceps exercises are done. Walking without a protective brace or corset is not permitted until the muscle is strong and the range of knee motion adequate. Recovery of muscle power may require about 6 months. The range of motion and strength is excellent, even without the patella.

JOINT DÉBRIDEMENT.[32, 33] When the joint is extensively involved and one wishes to relieve pain and preserve motion, the Magnuson operation is indicated. This consists of removal of all the accessible synovial membrane, osteophytes, diseased cartilage and abnormal soft tissues. It is necessary that the patient be capable of co-operating in the after-treatment which is quite painful and done immediately after operation.

Technic. A parapatellar incision is made, and the patella is displaced to one side. Magnuson recommends partial removal of the articulating portion of the patella, as much as one half of the bone. Complete removal may be indicated. It is probably wiser to remove it in all cases. The synovial membrane is dissected free and removed. Osteophytes are removed with a chisel. The degenerated cartilage is shaved down even to cancellous bone. The semilunar cartilages, if appearing to be healthy, may be left in situ. The cruciate ligaments should not be removed; otherwise, the ensuing instability will encourage further degeneration. When the patella is removed, the capsule in the quadriceps mechanism may require reinforcement with fascia lata strips or heavy braided silk. All loose bodies are cleaned out. Finally, the articular surfaces are smoothed down with a rasp or a file. Postoperatively, only a compression bandage is applied, and quadriceps setting exercises are instituted immediately. The range of motion improves steadily over a year.

ARTHRODESIS. Surgical obliteration of the joint provides absolute relief of pain and stability. It is indicated when destruction is extensive, when débridement has failed, and when rapid return to ambulation is desirable. When both knees are involved, preservation of motion in one knee is mandatory. Many methods for obtaining a fusion have been described, the multiplicity of procedures attesting to the difficulty and the uncertainty of accomplishing the arthrodesis. Postoperative immobilization is lengthy, and it is stated that most knees will fuse if immobilized long enough. However, failures as high as 40 per cent have been reported. The most successful

[32] Isserlin, B.: Joint débridement for osteoarthritis of the knee, J. Bone & Joint Surg. 32B:302, 1950.

[33] Magnuson, P. B.: Joint débridement; surgical treatment of degenerative arthritis, Surg, Gynec. & Obst. 73:1, 1941.

margins are enlarged, the range of active and passive motion is greatly diminished, and a fixed deformity in partial flexion is usual. Attacks of acute inflammatory synovitis accompanied by increased effusion and muscle spasm are common. Locking may be caused by nipping of a hypertrophied synovial fringe, displacement of an easily torn meniscus, or jamming of a detached osteophyte.

Pathology

The basic gross and microscopic findings of degenerative arthritis have been described in the section on Arthritis. In addition, certain conditions characterize knee-joint involvement. The patellofemoral articulation is the site of maximum damage. An ulcerative deepening of the undersurface of the patella corresponds to a site of maximal erosion over a femoral condyle. Many flakes of cartilage detached from the joint surface are found. Also, a variable number of rounded cartilaginous or osteocartilaginous loose bodies may represent stages in the continued growth of the flakes, or they may be formed by metaplasia in the synovium and be detached later. Degeneration also affects the menisci, making them extremely vulnerable to injury. Minute to gross tears with displacement are not uncommon Although participating in the joint deterioration, usually the cruciate ligaments do not succumb. They are generally found to be intact even in degeneration of extreme degree. The infrapatellar fat pad is thickened and somewhat indurated It may hypertrophy and protrude medially where it may be mistaken for a tumor.

Treatment

Conservative. Severe degenerative changes as seen in roentgenograms are compatible with a painless, stable, functioning knee provided that the quadriceps is strong and the joint is protected from unnecessary strains and pressures. Severe pain and effusion denote temporary irritation which frequently can be controlled by the following measures:

1. *Avoid weight-bearing*, if necessary by crutches.
2. *Immobilize the knee* by a cast, a splint, a knee corset, a brace or an elastic bandage.
3. *Heat*, preferably moist heat. Short-wave diathermy is destructive and should be avoided.
4. *Hydrocortisone* is injected at weekly intervals.
5. *Quadriceps exercises* are performed daily to maintain strength of the main stabilizing factor of the knee.
6. *Procaine needling* of a localized tender spot over the joint interval frequently relieves pain quickly.

The failure to respond to conservative treatment is sufficient indication for surgical intervention.

Surgical Treatment. Locking, irritation, pain and effusion are caused by an osteophyte, a large flake of cartilage, a hypertrophied synovial fringe, an enlarged fat pad, or a torn degenerated displaced meniscus Removal is indicated.

Patellectomy. Localized patellofemoral degenerative arthritis without involvement of the rest of the joint is an indication for removal of the patella. Failure to do this will eventuate in progression and damage to the entire knee. Tangential x-ray views of the patellofemoral articulation should be done routinely. The earliest signs of degenerative arthritis will be detected at this site, and patellectomy will prevent extension to the other joint surfaces.

Technic.[31] An incision is made skirting the medial or lateral border of the patella. The capsule about the patella, which is the continuation of the quadriceps tendon, is split longitudinally in the mid-line. The medial and the lateral sides of the patella are grasped with towel clips and displaced forward out of the wound. The patella is divided longitudinally through three fourths of its thickness by a hand saw, and section is completed manually or by a chisel. The two halves of the patella are spread, and the interior of the joint is inspected for loose bodies and other pathology. Next, the patellar halves are enucleated from the capsule by blunt dissection. The capsule is closed by imbrication. If the defect is weak, it can be reinforced by inserting strips of fascia lata or by sewing the quadriceps tendon to the patellar tendon by heavy gauge thread. Postoperatively, the knee is

[31] Boyd, H B, and Hawkins, B. L Patellectomy; a simplified technique, Surg, Gynec. & Obst 86 357, 1948.

tion in which the synovial membrane is congested, edematous and studded with tubercles. The joint fluid is increased, thin, watery, opalescent and contains flakes of fibrin and an increased number of mononuclear cells. Caseative destruction is almost nonexistent. On the contrary, the tendency is toward resorption of the tubercles or healing by fibrosis which thickens the synovium. If the inflammation persists and is more intense, it spreads to the periarticular tissues. The larger amount of granulation tissue is more likely to bridge the joint and obliterate the cavity with fibrous adhesions. The granulation tissue may invade the bone beneath the articular cartilage, which is loosened and extruded into the joint as the subchondral cortex is destroyed.

Extreme caseation destruction, abscesses and sinuses are uncommon. When it does occur, it is the result of spread of an acute exudative infection or focus of caseative necrosis in the metaphysis. This is a severe allergic reaction which causes greater intensity of inflammation in the bone (osteoporosis), joint (marked synovial swelling, increased effusion, abscesses, sinus formation) and periarticular tissues (marked swelling, heat, muscle spasm). Muscle atrophy of the thigh and the calf occur rapidly.

The usual low-grade synovitis *stimulates osteogenesis so that the ossification centers in the femur and the tibia are larger than on the opposite side.* As the infection continues, premature closure of the neighboring epiphyseal plate results in shortening of the extremity. In the early stage of muscle spasm, the hamstrings are chiefly affected and maintain the knee in flexion. With atrophy and contracture of these muscles and the posterior capsule, the flexion deformity becomes persistent. As the capsular apparatus softens, the tibia subluxates posteriorly. It is also abducted and rotated externally by the tight biceps femoris creating a valgus deformity.

Clinical Course, Symptoms and Findings at Various Stages

Infants and children below the age of 10 are predisposed. The onset is very insidious and often precipitated by trauma or an infectious disease such as measles. At first a *hydrarthrosis appears at intervals* and disappears, the joint being normal in appearance and function during each remission. Excessive weight-bearing activity and strains seem to precipitate each attack. The appearance is that of an ordinary synovitis with effusion distending the suprapatellar pouch, floating the patella, causing limitation only of the terminal arcs of motion and producing a slight limp and discomfort which is mild. No constitutional symptoms are present. The child otherwise seems to be healthy. The aspirated joint fluid is not abnormal except for the presence of a few mononuclears. At this stage the roentgenograms are negative.

Gradually, the attacks of synovitis become more intense and persistent, the synovium and the capsular structures are palpably thickened and generally tender, and the inflammation spreads to the periarticular tissues. The boggy swelling obliterates the patella and other bony prominences, stretches and whitens the overlying skin and produces the characteristic bulbous white swelling or *tumor alba.* At first, the muscles, particularly the hamstrings, are in spasm, then these undergo atrophy and contracture. The biceps femoris pulls the leg into a deformity of flexion, abduction and external rotation. If this deformity is allowed to remain, the tibia gradually subluxates posteriorly. Even when the inflammation reaches the most superficial structures, only a slight duskiness and mild tenderness are present. None of the characteristics of an acute inflammation, namely heat, redness, pain and marked tenderness, is apparent. The roentgenograms at this stage show generalized osteoporosis, loss of definition of the articular cortex, perhaps marginal erosions, increased density and outward bulging of the thickened synovium and capsule, and narrowing of the joint interval as the cartilage is destroyed. The ossification centers of the femur and the tibia may be enlarged. Aspirated fluid is thin, watery, opalescent, and contains large numbers of mononuclear cells and flakes of fibrin.

When healing takes place in the early stage of hydrarthrosis, full motion is restored. At a later period of intense diffuse inflammation, healing takes place by fibrosis and thickening of the capsule and formation of intra-articular adhesions causing a fibrous ankylosis.

An acute exudative or caseous infection in

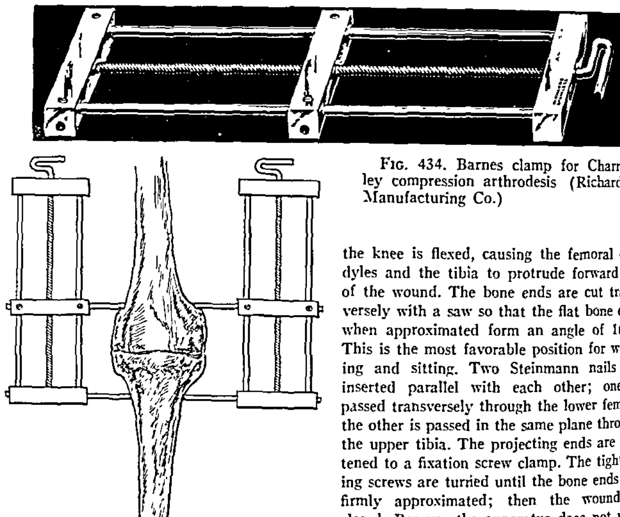


FIG. 434. Barnes clamp for Charnley compression arthrodesis (Richards Manufacturing Co.)

procedure is that which takes advantage of the principle of physiologic compression. Briefly, this concept states that two bony structures with adequate circulation when compressed together (the amount of pressure corresponding to normal muscular tension) will stimulate new bone formation and will unite. However, excessive pressure will cause the contacting surfaces to disintegrate. By the compression method fixation is rigid, the surfaces are firmly approximated, ambulation is early, and fusion is rapid and certain, occurring in 5 to 8 weeks.

Technic.[34, 35] Through a mid-line incision, the patellar tendon is severed at its insertion, and the patellar tendon, the patella and the quadriceps tendon are retracted upward. The patella is excised. The cruciates are cut, and the knee is flexed, causing the femoral condyles and the tibia to protrude forward out of the wound. The bone ends are cut transversely with a saw so that the flat bone ends when approximated form an angle of 160°. This is the most favorable position for walking and sitting. Two Steinmann nails are inserted parallel with each other; one is passed transversely through the lower femur, the other is passed in the same plane through the upper tibia. The projecting ends are fastened to a fixation screw clamp. The tightening screws are turned until the bone ends are firmly approximated; then the wound is closed. Because the apparatus does not prevent flexion and extension, the limb postoperatively is placed on a Thomas splint with the projecting pins resting on the lateral bars of the splint. Every few days the screws are tightened a little so that the pins are kept taut and slightly bent. After 4 weeks a cast is applied, incorporating the pins into the plaster, and ambulation with weight-bearing is permitted. At 8 weeks union is clinically firm. The pins may be removed at this time but the joint should be protected by a cast for an additional 4 weeks.

[34] Charnley, J. C Positive pressure arthrodesis of the knee joint, J. Bone & Joint Surg 30B 478, 1948

[35] Key, J. A Positive pressure in arthrodesis for tuberculosis of the knee joint, South M J 25 909, 1932.

TUBERCULOSIS OF THE KNEE JOINT

Tuberculous infection of the knee joint occurs typically in the infant or the child as *a low-grade synovitis* resulting in fibrotic thickening and a variable amount of restricted joint motion.

PATHOLOGY

A granular synovial infection *is the* most common type. This is a low-grade inflamma-

lesions elsewhere. (The exception of course is the uncommon joint infection complicating a severe osseous infection.) A period of conservatism is certainly warranted. If deformity or severe restriction of joint motion seems to be inevitable, or if a focus for future exacerbations remains, surgical excision and arthrodesis are the only means of permanent cure.

Treatment

Conservative. Streptomycin, PAS and INAH are given daily. The main principles in general treatment include absolute bed rest, heliotherapy, fresh air, a nutritious diet and good hygiene. The knee is immobilized by a cast which extends from the nipple line to the toes on the affected side. The cast is windowed about the knee. Once a week 1 Gm. of streptomycin is injected into the joint, and the intramuscular dose is withheld that day. A buffered alkaline solution is added to the articular injection to increase the effectiveness of the drug. The streptomycin is present in an effective level for 1 week in the joint fluid. A course consists of 3 months of treatment, during which time synovial effusion persists but subsides rapidly thereafter.[36] Traction is generally recommended for flexion and subluxation deformity. However, deformity is more likely in the more destructive infections wherein traction may be used as a preliminary to surgical arthrodesis.

Surgical Treatment. In an attempt to preserve motion, traction is first used to stretch the capsular structures. Next, tenotomy of the hamstrings and posterior capsulotomy aid in overcoming the deformity. Synovectomy and removal of intra-articular adhesions have been done, but improvement in motion is often inadequate. The danger of lighting up an infection or the possibility of future exacerbations argue against "conservative" surgery. Only osseous fusion is desirable. In performing arthrodesis in children the knee should be fused in full extension to compensate for future shortening and because some flexion occurs with further growth. Resection of bone should be sufficient to overcome deformity and to expose normal-appearing cancellous bone. The epiphyseal plates must be avoided. In adults the position of slight flexion is desirable for walking and sitting. In rare cases of severe destruction and draining sinuses, a mid-thigh amputation may be indicated as a lifesaving measure.

The technic of arthrodesis of the knee is described under "Degenerative Arthritis of the Knee." There is a variance of opinion as to whether or not weight-bearing is permissible as a *stimulus to fusion. At any rate,* immobilization should be prolonged and is not discontinued until trabeculations can be seen bridging the joint space.

BURSAE OF THE KNEE JOINT

As elsewhere, bursae about the knee function to reduce friction between surfaces. Although many bursae in this location have been described, a few are constant and of special significance:

1. **Suprapatellar bursa**—an upward extension of the joint synovium beneath the quadriceps tendon.
2. **Popliteal bursae**—the most important of these is that which is situated between the medial head of the gastrocnemius and the semimembranosus muscle.
3. **Prepatellar bursa**—lies anterior to the distal half of the patella.
4. **Infrapatellar bursa**—lies between the patellar tendon and the anterior surface of the tibia.
5. **Bursae of the collateral ligaments**—these small bursae lie beneath the tibial and the fibular collateral ligaments.
6. **Superficial pretibial bursa**—lies between the patellar tendon and the skin.
7. **Bursa anserina**—this is situated on the medial surface of the tibia deep to the tendons of the sartorius, the gracilis and the semitendinosus, and superficial to the insertion of the tibial collateral ligament.

Diseases of the Bursae

The synovium of the bursa is subject to pathology due to trauma, infection, metabolic disease, occupational strains and neoplasms.

1. **Acute Traumatic Bursitis.** Superficially situated bursae, such as the prepatellar and the pretibial bursae, are subject to direct trauma. The sac becomes distended with

[36] Stevenson, F H: The chemotherapy of orthopedic tuberculosis, J Bone & Joint Surg 36B:5, 1954.

the metaphysis of the femur or the tibia may produce irritation of the synovium, a nonspecific synovitis. When the infection actually invades the joint, inflammatory signs are sudden and more intense and cause severe pain, muscle spasm, heat, tenderness, and great restriction of motion. Destruction is greater, and abscess and sinus formation more frequent. Constitutional symptoms include afternoon fever, anorexia, night sweats, etc. This type is more common in the adult.

Secondary Changes. Destruction causes shortening. Premature fusion of the epiphyseal line results in retardation of longitudinal growth. Stimulation of ossification of an epiphysis occurs notably in the medial femoral condyle, producing bony enlargement and a valgus deformity. The characteristic flexion, abduction and external rotation are due to a tight biceps. Posterior subluxation occurs in late neglected cases

Laboratory Diagnosis

Roentgenographic. *The roentgenograms* within the early weeks are negative. When tuberculosis is suspected and x-ray studies are unrevealing, these should be repeated at regular intervals. The following features may eventually appear.

A. *Osteoporosis* of juxta-articular structure is the earliest finding. Its earlier appearance and intensity of decalcification are proportionate to the severity of the infection.

B. *Ballooning of the capsule.* Fibrosis increases the capsular density. The lines of the capsule are bulged outward by the effusion.

C. *Swelling of periarticular soft tissue.*

D. *Enlargement of ossification centers.* The ossification centers adjacent to the affected joint appear earlier and enlarge more rapidly than do those of the opposite knee. The epiphyseal plates tend to close prematurely

E. *Loss of definition of articular cortex.* The articular cortices become extremely thinned but the marginal contours are well preserved in the early stages When the infection is of long standing and invades the subarticular bone, marginal erosions become evident. With further destruction, the bone ends become ragged.

F. *Narrowing of joint interval.* This is a late finding. As the subarticular bone is invaded from the synovium or, as in adults, by spread from a metaphyseal focus, the articular cartilage becomes separated and extruded into the joint cavity. In consequence, the epiphyses are more closely apposed.

G. *Osseous lesions.* Usually occurring in adults, the metaphyseal involvement takes several forms. Intense diffuse osteoporosis signifies an acute exudative process, and the adjoining hydrarthrosis constitutes an irritative synovitis. When this lesion heals, the original trabecular architecture is completely restored. Or multiple small or slightly larger circumscribed foci of osteolysis may be observed in an osteoporotic metaphysis. These granular foci may fuse into a single large defect. Healing takes place by the appearance of an encircling ring of dense bone which encroaches upon, narrows and obliterates the defect. Failure of healing and progressive destruction is evidenced by complete disintegration of the osseous structure. Densities of necrotic bone sequestra occupy large ragged osteolytic defects. The articular margin becomes ragged and eventually the opposite articular cortex is secondarily involved. A clear osteolytic cavitation within the bone may display gradually increasing density at its center. This phenomenon is caused by the deposition of calcium salts in caseous material.

H. *Deformity.* Posterior subluxation of the tibia is characteristic of advanced cases. A valgus deformity may occasionally be produced.

Biopsy. Microscopic examination of a synovial specimen yields the highest percentage of positive results.

Culture and Guinea Pig Inoculation. These require many weeks to perform and give less accurate results. However, these tests are done for confirmation. It must be remembered that the fluid in a nonspecific synovitis reacting to a neighboring tuberculous infection is sterile.

Prognosis

In the early stage of hydrarthrosis conservative treatment will often result in excellent restoration of joint function. Typically, these patients do not have multiple or severe

and a compression bandage is applied. A new bursa will re-form.

3. **Acute Infectious Bursitis.** A penetrating wound implants organisms within the cavity. The synovium is severely congested and swollen. Exudate, which distends the sac, is serosanguineous in streptococcal infection; purulent, in staphylococcus infection. Clinically, swelling is rapid, very painful and exquisitely tender. The overlying skin is reddened and warm. Muscle spasm limits joint motion. Constitutional reaction is evidenced by fever, leukocytosis and an increased sedimentation rate.

Treatment. If the infection seems to be mild, the exudate may be aspirated, and an antibiotic injected. The knee is splinted, and hot packs are applied. More often, incision is required. A drain is inserted, and the patient is positioned to secure dependent drainage. Antibiotics are administered, but sensitivity tests will reveal the most effective one.

4. **Chronic Infectious Bursitis.** The prepatellar bursa is a favored site for *tuberculous* infection. Swelling develops very gradually, and pain and tenderness are moderate. The overlying skin is thickened but lacks the redness and the warmth of an acute infection. The course is prolonged. Diagnosis is mainly by biopsy.

The prepatellar bursa is involved in the tertiary stage of syphilis. Swelling develops slowly and is doughy to palpation. Pain, tenderness and local inflammatory signs are characteristically absent. The overlying soft tissues may disintegrate, and a chronically draining sinus will result. Diagnosis is established by biopsy and serology.

Treatment. A tuberculous bursa is completely excised, and the knee is immobilized for a long period of time. Streptomycin, PAS and isoniazid are administered. For the syphilitic bursitis, antiluetic therapy is effective.

5. **Gouty Bursitis.** Urate deposits in the bursal wall cause local symptoms and signs comparable with an acute infectious process. The prepatellar bursa is often involved by extension from the patella, although other bursae may be affected similarly. Eventually, the bursal walls become thickened and fibrotic. Biopsy and microscopic examination of aspirated fluid reveal the typical urate crystals.

Treatment. The thickened, tender gouty bursa and the underlying bony lesion may be excised. An antigouty program is instituted. (See section on "Affections of Joints.")

6. **Bursitis Associated With Arthritis.** A bursa communicating with the joint cavity, e.g., the popliteal bursa, is affected by an extension of the same pathologic process. Acute symptoms of arthritis are associated with concomitant symptoms and swelling of the communicating bursa.

Treatment. Management of rheumatoid and osteoarthritis will control the bursitis effectively. If the bursa is thickened and the site of chronic inflammation, giving rise to persistent symptoms, it should be extirpated.

7. **Neoplasms of Bursae, Osteochondromatosis, Villonodular Synovitis, Xanthomatosis and Synovioma.** These may involve the bursa independently or as an extension of a similar lesion in the communicating knee joint. The popliteal bursa is a favored site for fibrosarcoma.

Treatment. Excision of the bursa is recommended for all but malignant lesions which require amputation and irradiation.

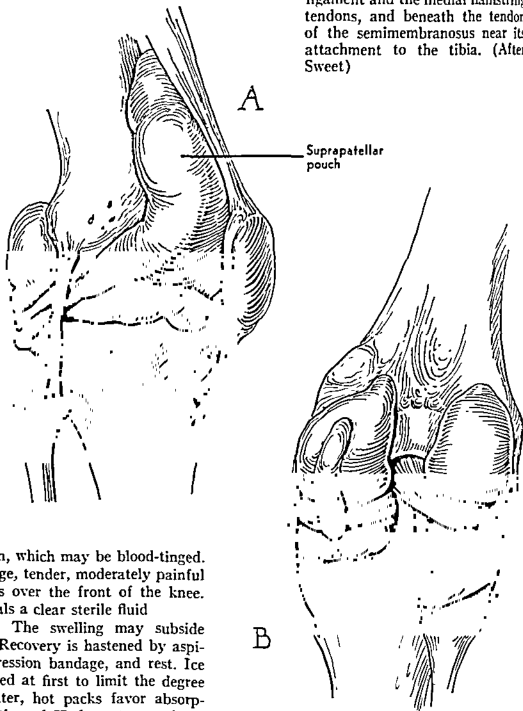
8. **Calcified Bursitis.** Heterotopic amorphous calcium deposits are rarely observed. The cause is unknown. It may be an extension from an adjacent calcified tendinitis similar to that seen in the shoulder.

BURSITIS IN SPECIAL SITUATIONS

Popliteal Bursae

Numerous and variable bursae have been described in relation to the hamstring tendons, the heads of origin of the gastrocnemius, the posterior capsule, and the femoral and the tibial condyles. The most important of these are: (1) the bursa anserina which lies between the tendons of the sartorius, the gracilis and the semitendinosus superficially and the tibia and the tibial collateral ligament deeply; (2) a semimembranosus bursa between the tibial collateral ligament and the tendon of insertion of the semimembranosus; (3) a second semimembranosus bursa located between the semimembranosus tendon and the medial tibial condyle; (4) a third semimembranosus bursa situated between the medial head of the gastrocnemius and the overlying semimembranosus muscle; (5) a bursa lying between the

FIG. 435. Synovium and common bursae about the knee joint. (A) Lateral view. The suprapatellar extension of the synovial pouch is the site of accumulation of effusions. Not shown are the suprapatellar and the superficial infrapatellar bursae. Note that the synovial pouch extends behind the femoral condyles. (After Spalteholz) (B) Posterior view. The elongated bursa seen at the left side of the figure lies between the medial head of the gastrocnemius and the semimembranosus. It is often the site of a chronic bursitis. Not shown are the bursae between the fibular collateral ligament and the biceps tendon, between the tibial collateral ligament and the medial hamstring tendons, and beneath the tendon of the semimembranosus near its attachment to the tibia. (After Sweet)



synovial effusion, which may be blood-tinged. Clinically, a large, tender, moderately painful swelling appears over the front of the knee. Aspiration reveals a clear sterile fluid

Treatment. The swelling may subside spontaneously. Recovery is hastened by aspiration, a compression bandage, and rest. Ice packs are applied at first to limit the degree of swelling. Later, hot packs favor absorption. The injection of Hydrocortone reduces the swelling quickly.

2. **Chronic Traumatic Bursitis.** Repeated traumata, such as that sustained in occupations requiring kneeling, causes recurring effusions. The bursal wall becomes thickened and fibrotic. Villi, folds and bodies, which are small accumulations of fibrin, may form within the synovial cavity. A persistent tender and painful swelling is present.

Treatment. The sac is completely excised,

through a curved incision which parallels one border of the patella. The superficial wall of the bursa is often densely adherent to the skin.

Infrapatellar Bursitis. This bursa is situated between the upper anterior surface of the tibia and the patellar tendon. When swollen, it limits flexion and extension of the knee joint. Swelling and tenderness are perceived on either side of the patellar tendon.

Treatment. If swelling is not reduced by heat, splinting, aspiration, and injection of Hydrocortone, the bursa must be excised.

Anserina Bursitis. Effusion and thickening are due to repeated minor traumata. A painful swelling is discerned over the upper and medial aspect of the tibia.

Treatment. Heat, splinting, aspiration, and injection of hydrocortone usually effect a cure. Persistent swelling should make one suspicious of other pathology. Excision is advisable.

inner head of gastrocnemius and the medial femoral condyle; (6) a rare bursa between the tendons of the semimembranosus and semitendinosus tendons.[37]

Often the bursa beneath and that overlying the inner head of the gastrocnemius fuse and form a composite bursa, designated *the gastrocnemio-semimembranosus bursa*. Many of these composite bursae communicate with the knee joint through an opening in the posterior capsule located superior and medial to the oblique popliteal ligament and directly beneath the most proximal portion of the inner head of the gastrocnemius. The bursa superficially is lax and loosely attached to the areolar tissue. Deeply, the bursa fuses intimately with the tendon of the semimembranosus and the inner head of the gastrocnemius. Then it is reflected on the posterior capsule.

A popliteal cyst is a distention of a popliteal bursa, most commonly the gastrocnemio-semimembranosus bursa. It is commonly known as a *Baker's cyst*. The very probable cause is the repeated traumata due to constant friction between adjacent muscles. Another theory postulates a herniation of synovium through a defect in the posterior capsule.

Clinical Picture

All ages are affected, males predominating. It may be bilateral. A cystic, tense, painless, mobile swelling appears gradually in the popliteal fossa, usually over the medial side It is rendered more prominent by extending the knee joint. Occasionally, vague symptoms of pain and a sense of giving-way are complained of. If the mass is large, full extension of the joint is limited. On palpation, no tenderness can be elicited. Fluctation may be detected. The overlying skin is not adherent. Aspiration reveals a clear, viscid fluid.

Roentgenologic Findings

A globular shadow is observed in the popliteal fossa having a greater density than surrounding soft-tissue structures. Air arthrography of the knee joint often demonstrates a communication with the sac.

[37] Wilson, P. D, Eyre-Brook, A L, and Francis, J D A clinical and anatomical study of the semimembranosus bursa in relation to popliteal cyst, J. Bone & Joint Surg 20 963, 1938

Differential Diagnosis

Other causes of swellings in the popliteal fossa include:

1. **Aneurysm**—pulsating tumor, bruit synchronous with the pulse, arteriography
2. **Hemangioma**—painful, tender, soft swelling. Increased local temperature. Superficial vessels dilated. Multiple opacities of calcification observed in roentgenograms.
3. **Neoplasms**—benign and malignant. Benign tumors are soft, mobile and well-localized. Malignant fibrosarcoma is painful, firm, tender, and fixed to surrounding tissues. The regional lymph nodes may be involved. Diagnosis is established by biopsy.

Treatment

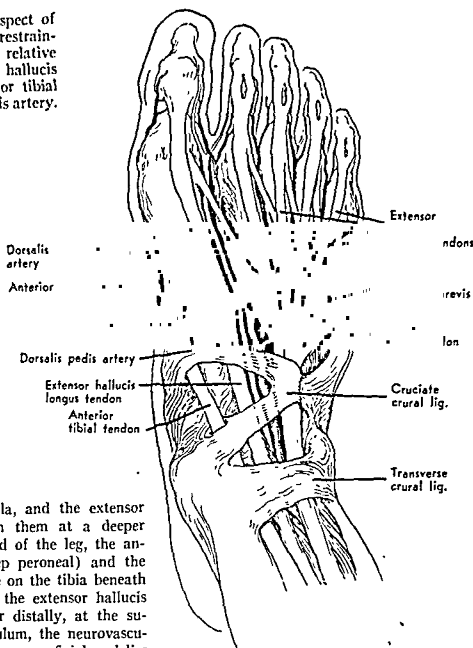
The popliteal cyst must be excised if it is productive of symptoms or for cosmetic reasons.

Technic. An incision is made longitudinally over the medial femoral condyle, then turns transversely at the level of the joint line to the middle of the fossa and is extended a little distally. As the deep fascia is incised, the cyst bulges into view. The superficial portion of the sac is freed by sharp dissection. The deep portion between the semimembranosus and the inner head of the gastrocnemius is adherent to the tendinous surfaces of these muscles and is freed by cutting away a thin layer of the tendinous tissue. The neurovascular structures in the popliteal fossa are protected by retracting the inner head of the gastrocnemius to the lateral side. Then the neck or the base of the cyst is approached where it enters the posterior capsule. It is severed near the attachment. It is unnecessary to close the opening in the capsule. Postoperatively, a compression bandage is applied.

Prepatellar Bursitis. Chronic distention of the prepatellar bursa occurs as a result of repeated trauma in kneeling which certain occupations entail. It is known as "housemaid's knee." The walls become thickened, and the interior is loculated by irregular adhesions and septa. Loose rice-bodies are often found.

Treatment. Spontaneous subsidence may occur if pressure against the front of the knee is eliminated If the demands of an occupation must be served, a sponge rubber knee pad is worn. Otherwise, the bursa is excised

FIG. 436. The dorsal aspect of the foot. Note the fascial restraining bands. Note also the relative positions of the extensor hallucis longus tendon, the anterior tibial nerve and the dorsalis pedis artery.



laterally along the fibula, and the extensor hallucis longus between them at a deeper level. In the lower third of the leg, the anterior tibial nerve (deep peroneal) and the anterior tibial artery lie on the tibia beneath the anterior tibial and the extensor hallucis longus muscles. Further distally, at the superior extensor retinaculum, the neurovascular bundle becomes more superficial and lies between the extensor hallucis longus and the extensor digitorum longus. It then passes into the foot under the inferior extensor retinaculum. The artery throughout its course at this level lies immediately lateral to the anterior tibial nerve. About 5 cm. above the ankle joint, the anterior tibial artery gives off 2 branches, the anterior medial malleolar and the anterior lateral malleolar arteries, which run medially and laterally, respectively, to join an arterial network about the malleoli. The lateral anterior malleolar artery anastomoses with the perforating branch of the peroneal behind the extensor digitorum longus. These arteries form an extensive network over the anterior aspect of the ankle joint and must be dealt with in operative

Tibialis Anterior Muscle. This tendon lies against the lateral surface of the tibia. It inserts into the medial aspect of the first cuneiform bone and the adjacent base of the first metatarsal. It is a dorsiflexor and invertor of the foot.

Extensor Digitorum Longus Muscle. The tendon lies immediately medial to the fibula, and in passing through the inferior retinaculum it divides into 4 tendons which insert on the middle and the distal phalanges of the lateral 4 toes. On the dorsum of the first phalanx of the 2nd, the 3rd and the 4th toes each tendon is joined on its lateral side by a tendon from the extensor digitorum brevis to form an expansion over the toe. Each expansion is composed of a middle slip which inserts

26

The Foot and the Ankle

SURGICAL ANATOMY

The function of the ankle and the foot is so interrelated that their surgical anatomy should be considered together.

Cutaneous Veins

Two *dorsal digital veins* of each toe join to form a common stem which with the digital veins of other toes form the *dorsal venous arch*; this arch lies in the superficial fascia over the distal ends of the metatarsals. The medial end of the dorsal venous arch joins the medial dorsal digital vein of the big toe to form the *long saphenous vein*, which ascends in front of the medial malleolus into the leg. The lateral end of the dorsal venous arch unites with the lateral dorsal digital vein of the little toe to form the *short saphenous vein*, which passes backward below the lateral malleolus and ascends into the leg.

Cutaneous Nerves

The medial side of the dorsum of the foot is supplied by the *saphenous nerve*, the lateral side by the *sural nerve*, and the intermediate area by the *musculocutaneous nerve*. The musculocutaneous nerve arises from the common peroneal at the neck of the fibula and descends under cover of the peroneus longus muscle. It becomes superficial at the distal third of the leg and divides into 2 branches, which supply the intermediate portion of the dorsum of the foot before supplying the toes. The sural nerve arises from the posterior tibial nerve, enters the foot below the lateral malleolus, runs along the lateral border of the foot along the short saphenous vein and supplies the lateral border of the little toe.

Deep Fascia

The deep fascia over the lower leg is incomplete over the tibia and the fibula, where it attaches to the margins of the subcutaneous aspects of those bones. It is very thin over the distal end of the leg and the foot except where it forms thick bands which act as slings or pulleys to bind down the tendons and provide an efficient mechanical structure for strong pull by the muscles.

The *superior extensor retinaculum* (transverse crural ligament) is a broad band which stretches across the front of the leg above the ankle from the tibia to the fibula. Tendons transferred to the front of the leg must be routed beneath the retinaculum.

The *inferior extensor retinaculum* (cruciate crural ligament) crosses the dorsum of the foot. It is attached medially by 2 bands: one to the medial malleolus, the other to the deep fascia of the sole. Laterally, it is fixed to the anterior part of the calcaneus. These retinacula strap down the anterior tibial, the extensor hallucis longus and the extensor digitorum longus.

A fascial band, the *superior peroneal retinaculum*, binds down the peronei longus and brevis on the back of the lateral malleolus Below this, the *inferior peroneal retinaculum* hold these tendons to the lateral surface of the calcaneus.

Peritendinous Synovial Sheaths

Each of the 3 tendons—the tibialis anterior, the extensor hallucis longus and the extensor digitorum longus—are surrounded by synovial sheaths where they pass beneath the retinacula on the dorsum of the foot. The sheath of the tibialis anterior extends as far as the big toe. Similarly, the peronei are enveloped by synovium behind and below the lateral malleolus. This is a favorite site for tuberculous infection, which can extend along the peroneus longus to the tarsal joints

Relationships of Structures in the Anterior Compartment of the Leg Above the Ankle

The anterior tibial muscle lies medially along the tibia, the extensor digitorum longus

Peroneal Tendons. These tendons descend behind the lateral malleolus, the longus lying superficial to the brevis, and are bound down by the superior peroneal retinaculum. They then curve downward and forward, bound to the lateral surface of the calcaneus by the inferior peroneal retinaculum. The superior peroneal retinaculum may be cut in the approach to the lateral ligaments of the ankle without fear of dislocation of the tendons, because they are still retained in place by the inferior retinaculum. Distally, the peroneus brevis comes to lie superficial to the longus on its way to its insertion into the 5th metatarsal bone. The peroneus longus passes into the sole beneath the cuboid bone to reach its insertion at the base of the 1st metatarsal bone. These muscles act as evertors and partially as plantar flexors of the foot. The peroneus tertius has been described above.

Musculocutaneous Nerve. This branch of the common peroneal nerve descends into the substance of the peroneus longus, then in the lower leg lies between the brevis and the longus, crosses over the brevis to reach the anterior aspect of the ankle, which it supplies, and passes distally to supply, by 2 terminal branches, the intermediate portion of the dorsum of the foot and most of the toes.

Relationships of Structures Over the Posterior Aspect of the Ankle

The deep fascia is thin except on the lateral side of the ankle, where it is thickened to form the superior peroneal retinaculum, and over the medial side, where a band bridges the interval between the calcaneus and the medial malleolus, the *flexor retinaculum*. The latter straps down the flexors and the tibialis posterior tendons. The most superficial structure beneath the deep fascia is the Achilles tendon, the largest tendon in the body, which arises from the gastrocnemius and the soleus muscles. It is attached to the middle of the posterior surface of the calcaneus. A small bursa separates the tendon from the upper

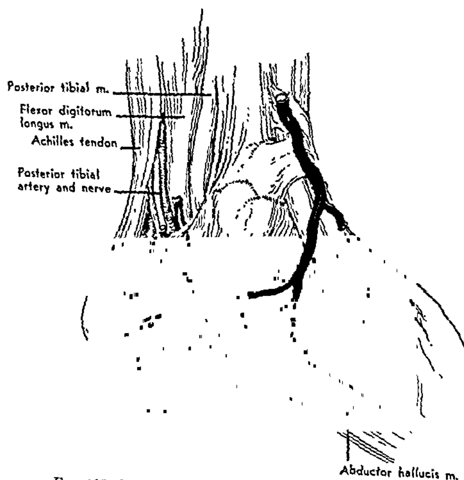


Fig. 438. Structures of the medial aspect of the ankle.

into the base of the middle phalanx and 2 lateral slips which are joined by tendons from the lumbrical and the interossei to insert into the base of the distal phalanx. The little toe has only one extensor tendon aside from the intrinsics. The extensor digitorum longus acts as an extensor of the toes and a dorsiflexor of the foot.

Peroneus Tertius Muscle. This small inconstant muscle arises from the distal anterior surface of the fibula, joins the extensor digitorum longus and is inserted into the dorsal aspect of the base of the 5th metatarsal bone. It is a dorsiflexor of the foot and an evertor of the foot.

Extensor Hallucis Longus. The tendon at the level of the ankle lies between the tibialis anterior and the extensor digitorum longus. From this point distalward it lies medial to the anterior tibial nerve and the dorsalis pedis artery. It inserts into the base of the distal phalanx of the large toe, acting as an extensor of the toe and a dorsiflexor of the foot.

Anterior Tibial Nerve. This branch of the common peroneal, also known as the deep peroneal nerve, lies lateral to the anterior tibial artery at the ankle and the dorsalis pedis in the foot. At the level of the talus upon which it lies, it divides into 2 terminal branches. The medial branch continues forward to supply the adjacent aspects of the big toe and the second toe. The lateral branch turns abruptly laterally and ends in a gangliform enlargement on the dorsum of the talus beneath the extensor digitorum brevis muscle

Anterior Tibial Vessels. This artery is accompanied by 2 venae comites and at the level of the ankle becomes known as the *dorsalis pedis artery.* The latter runs alongside the anterior tibial nerve and its medial terminal branch to the proximal part of the first interosseous space where it dips plantarward to join the lateral plantar artery in forming the plantar arch. It gives off small tarsal arteries and the first dorsal metatarsal artery. The dorsalis pedis artery at its distal end turns laterally and continues across the foot as the *arcuate artery.* This arterial arch sends 3 *dorsal metatarsal arteries* forward to the lateral 3 web spaces where each divides into 2 *dorsal digital arteries,* one for each of 2 contiguous toes. The first dorsal metatarsal artery divides into dorsal digital arteries for the medial side of the big toe and the adjacent aspects of the big and the second toes.

Extensor Digitorum Brevis Muscle. This muscle originates from the dorsal aspect of the calcaneus and sends 4 tendons to join the tendons of the extensor digitorum longus and the extensor hallucis at the base of the big, the 2nd, the 3rd and the 4th toes. It assists in extending the middle and the distal phalanges In approaches to the tarsal area, this muscle is reflected from its origin.

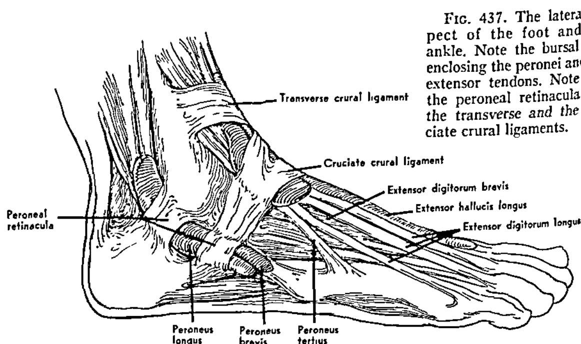


FIG. 437. The lateral aspect of the foot and the ankle. Note the bursal sacs enclosing the peronei and the extensor tendons. Note also the peroneal retinacula and the transverse and the cruciate crural ligaments.

cover of the flexor hallucis longus. It gives off a perforating branch, near the inferior tibiofibular joint, which pierces the interosseous membrane and descends over the lower part of the fibula to the dorsum of the foot. The peroneal artery runs behind the peroneal tendons and breaks up into a number of lateral calcaneal branches.

Flexor Hallucis Longus Muscle. The origin is the posterior surface of the fibula. Its tendon occupies a deep groove on the posterior surface of the talus, then passes into the foot beneath the flexor retinaculum, runs along the sole and is inserted into the base of the terminal phalanx of the large toe. It is a plantar flexor of the large toe and assists in plantar flexion and inversion of the foot.

Flexor Digitorum Longus Muscle. This muscle arises from the posteromedial surface of the tibia. Its tendon enters the sole of the foot under cover of the flexor retinaculum and divides into 4 tendons, each of which is inserted into the terminal phalanx of the lateral 4 toes. It flexes these toes and acts as a plantar flexor and invertor of the foot.

Tibialis Posterior Muscle. This muscle arises from the interosseous membrane and the posterior surfaces of the tibia and the fibula. The tendon runs toward the flexor retinaculum under cover of the flexor digi-

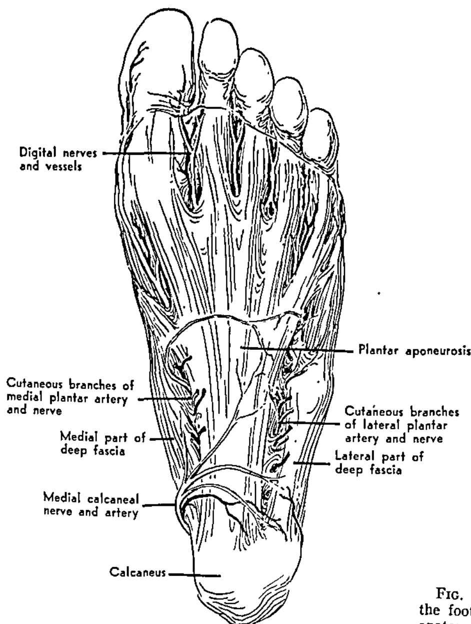


FIG. 440. Sole of the foot, superficial anatomy.

part of the posterior surface of the calcaneus.

Beneath the Achilles tendon are seen the tendons and the neurovascular bundle which pass obliquely toward and below the medial malleolus. The most lateral structure is the flexor hallucis longus tendon as it issues from the muscle which is attached to the fibula.

Under the fibular attachment lies the peroneal artery which descends along the fibular shaft, gives off a perforating branch above the ankle and passes below the lateral malleolus to run along the lateral surface of the calcaneus. Immediately lateral to the flexor hallucis longus tendon is the neurovascular bundle Lateral to the bundle is the tendon of the flexor digitorum longus beneath which lies the tendon of the tibialis posterior.

All these structures are held deeply by a thin layer of investing fascia As the tendons, the nerves and the vessels pass toward the medial border of the foot beneath the flexor retinaculum, they lie in the following order from front to back: tendon of the tibialis posterior, tendon of the flexor digitorum longus, end of the posterior tibial artery and the beginning of the medial and the lateral plantar arteries, with their venae comites, posterior tibial and medial and lateral plantar nerves, and tendon of the flexor hallucis longus

The flexor retinaculum holds these structures close to the medial malleolus. Any incision over the medial aspect of the os calcis must be placed an inch below the malleolus

Posterior Tibial Nerve. This nerve, after supplying the above-named muscles and the deeper part of the soleus in the upper leg, sends cutaneous twigs which pierce the flexor retinaculum and supply the skin over the posterior and the lower surfaces of the heel. Articular branches supply the posterior capsule of the ankle joint.

Posterior Tibial Artery. This is one of the two terminal branches of the popliteal artery. Under cover of the flexor retinaculum it divides into medial and lateral plantar arteries It sends a communicating branch which runs laterally to join the peroneal artery.

Peroneal Artery. This artery arises from the posterior tibial just below the knee and passes obliquely outward, then runs distally ; the medial aspect of the fibula under

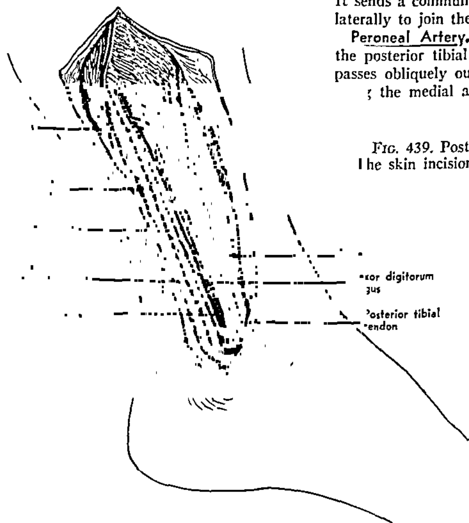


FIG. 439. Posteromedial aspect of ankle. The skin incision lies one finger's breadth behind the medial malleolus. The deep fascia has been incised. A deeper layer is seen to extend transversely across the leg interposed between the Achilles tendon and the deeper structures, binding the latter anteriorly against the tibia. The deeper fascial sheath is incised over the flexor hallucis longus tendon, which is palpable in the deeper and lateralmost portion of the wound where it lies over the fibula.

interphalangeal joints. The 2 slips also pass around the metatarsophalangeal articulations to attach to the transverse metatarsal ligaments which bind the metatarsal heads together. Opposite the webs between the toes, in the intervals between the processes, the digital vessels and nerves and the lumbrical tendons are seen. At the lateral border of the central portion of the deep fascia, 2 strong vertical intermuscular septa are passed upward on either side of the flexor digitorum brevis muscle. The borders of the central portion are continuous with the lateral and the medial portions. The medial portion of the deep fascia encloses the abductor hallucis muscle. The lateral portion encloses the abductor digiti quinti muscle. The deep fascia, particularly its central portion, is known as the *plantar aponeurosis*.

The muscles of the sole are disposed in 4 layers. The first layer consists of the short flexor of the toes and the abductors of the big and the little toes.

Flexor Digitorum Brevis Muscle. This muscle originates from the medial tubercle of the calcaneus and passes distally in the central part of the sole to the metatarsal area where it divides into 4 slips which send tendons inserting into the middle phalanx of the toes. Opposite the first phalanx, each tendon divides into 2 slips to permit passage of the long flexor tendon. It acts to flex the first interphalangeal joint. On either side of the muscle, the medial and the lateral plantar arteries and nerves make their appearance in the sole.

Abductor Hallucis Muscle. The flexor retinaculum and the medial tubercle of the calcaneus give origin to this muscle. The tendon joins the medial tendon of the flexor hallucis brevis to insert into the base of the proximal phalanx of the big toe. Its function is to abduct the big toe from the second toe.

Abductor Digiti Quinti. This muscle arises from both tubercles of the calcaneus The tendon is inserted into the lateral side of the proximal phalanx of the little toe. It abducts the small toe from the 4th toe.

Medial Plantar Nerve. This nerve originates from the posterior tibial nerve under cover of the flexor retinaculum. It becomes superficial in the sole in the interval between the flexor digitorum brevis and the abductor hallucis muscles. It gives off a large digital branch to the medial side of the large toe. Then it sends forward a digital nerve toward the first 3 webs, where each divides into 2 branches to supply adjacent aspects of 2 toes. Thus the medial plantar nerve supplies the skin over the plantar aspect of the 1st, the 2nd, the 3rd and half of the 4th toes. The most lateral digital nerve sends a communicating branch to the lateral plantar nerve.

Muscular branches go to the abductor hallucis, the flexor digitorum brevis, the flexor hallucis brevis and the 1st lumbrical.

Lateral Plantar Nerve. The lateral plantar nerve arises from the posterior tibial under cover of the flexor retinaculum and becomes superficial in the sole in the interval between the flexor digitorum brevis and the abductor digiti quinti. It gives off branches to the abductor digiti quinti and the flexor digitorum accessorius. It then sends a digital nerve to the lateral side of the little toe and another to the outer web space, where it divides into 2 branches to supply the adjacent surfaces of the 4th and the 5th toes. At this point the 3rd plantar and the 4th dorsal interosseous muscles and the flexor digiti quinti brevis receive their nerves of supply. The lateral plantar nerve then sends a deep branch which proceeds medially, like the deep branch of the ulnar in the palm. It supplies the adductor of the big toe, the lateral 3 lumbricals, the remainder of the interossei, and the intertarsal and the tarsal-metatarsal joints.

Medial Plantar Artery. This is the smaller of 2 terminal branches of the posterior tibial artery which arise beneath the flexor retinaculum. It appears between the flexor digitorum brevis and the adductor hallucis At the base of the first metatarsal it anastomoses with the first dorsal metatarsal artery, then passes along the medial border of the big toe, where it is joined by a digital branch from the first plantar metatarsal artery.

Lateral Plantar Artery. This artery runs across the sole to the base of the 5th metatarsal bone, where it turns medially with the deep branch of the lateral plantar nerve. This second portion of the artery is known as the *plantar arch*, which ends at the proximal end of the first metatarsal space by joining the end of the dorsalis pedis artery. The plantar arch lies on the bases of the metatarsals and

torum longus. It is inserted into the tuberosity of the scaphoid, the inner cuneiform bone, and by slips to other bones of the foot. Its function is to plantar flex and invert the foot.

The Sole of the Foot

The skin is very thick over the heel, the ball and the lateral border of the foot. The superficial fascia is a thick, tough membrane which is intimately attached to the overlying skin by fibrous bands which subdivide the subcutaneous fat into lobules. The deep fascia is a thick, strong membrane consisting of pearly white glistening fibers longitudinally disposed. It may be divided into central, medial and lateral portions. The central portion is thickest. Its narrow posterior end attaches to the medial process of the tuberosity of the calcaneus. Distally, it becomes broader and thinner and divides opposite the metatarsophalangeal joints into 5 processes—one for each toe. At the point of division, numerous transverse fasciculi strengthen the fascia. Each process to the toe divides into a superficial and a deep layer. The superficial layer inserts into the transverse sulcus of the skin which separates the toe from the sole. The deep layer separates into 2 slips which embrace the flexor tendons and are continuous with fibrous sheaths which envelop the tendons over the phalanges but not over the

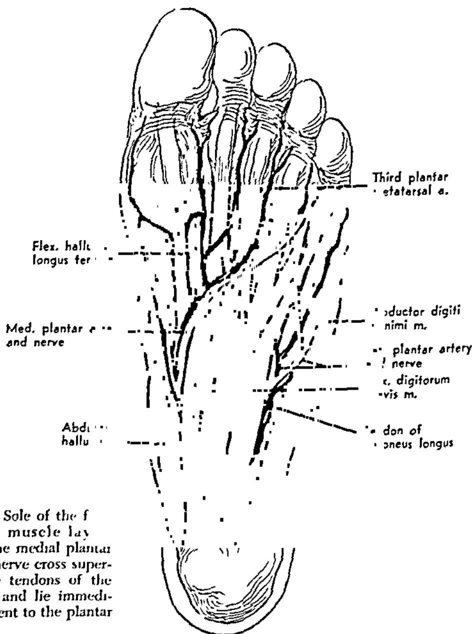


Fig. 441. Sole of the f superficial muscle la Note that the medial planta artery and nerve cross superficial to the tendons of the short flexor and lie immediately subjacent to the plantar aponeurosis.

FIG. 443. Sole of the foot, deep muscle layer. The tendons of the flexor digitorum longus have been removed.

muscle flexes the first metacar joint. Each tendon contains a se which lies in relation to and art the head of the 1st metatarsal.

Adductor Hallucis Muscle. T. composed of 2 heads, an oblique and a transverse. The oblique head arises from the fibrous sheath of the peroneus longus and the bases of the 2nd, the 3rd and the 4th metatarsal bones The transverse head arises from the plantar ligaments of the lateral 4 metatarsophalangeal joints. The 2 heads converge and by a common tendon insert with the lateral tendon of the short flexor into the lateral aspect of the base of the proximal phalanx. The oblique head adducts and flexes the big toe. The transverse head draws the roots of the toes closer together and accentuates the metatarsal arch.

Flexor Digiti Quinti Brevis Muscle. The short flexor of the little toe springs from the base of the 5th metatarsal and the fibrous sheath of the peroneus longus. The muscle belly envelops the metatarsal shaft and inserts into the lateral side of the base of the proximal phalanx. It flexes the metatarsophalangeal joint.

The lateral plantar nerve may be considered, along with the plantar arterial arch, as lying in the third layer; the deep division of the nerve runs beneath the oblique head of the short flexor of the big toe across the sole and lies upon the interosseous muscles and the metatarsal bones. The nerve lies immediately behind the plantar arch.

Plantar Arch. The plantar arch is a continuation of the posterior tibial artery, originating at the base of the 5th metatarsal bone and receiving the dorsalis pedis artery through the proximal portion of the first interosseous space. It curves across the sole in front of the deep branch of the lateral plantar nerve. Three perforating branches pass through the lateral 3 metatarsal spaces to join the corresponding dorsal metatarsal arteries. It sends

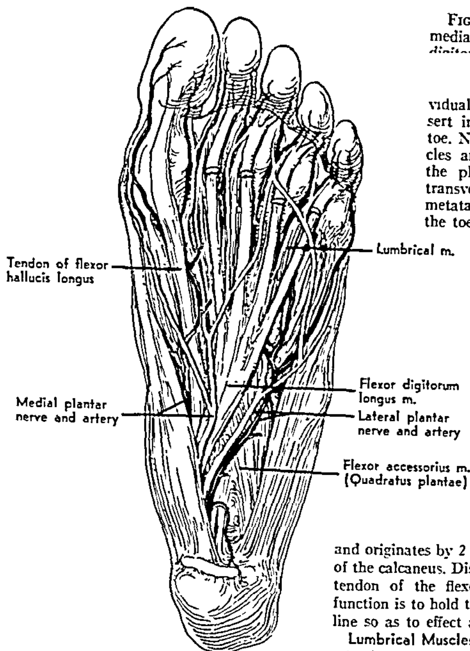


FIG. 442. Sole of the foot, intermediate muscle layer. The flexor digitorum b...

vidual tendons each of which insert into the distal phalanx of a toe. Note that the lumbrical muscles and digital nerves run over the plantar aspect of the deep transverse ligaments (between the metatarsal heads) on their way to the toes.

the oblique head of the flexor hallucis brevis. It gives off perforating branches which penetrate the 2nd, the 3rd and the 4th interosseous spaces and anastomose with the dorsal metatarsal arteries It sends 4 plantar metatarsal arteries forward, which divide at the web into plantar digital arteries—one for each toe.

The second layer of muscles consists of the tendon of the flexor hallucis longus, and those of the flexor digitorum longus and the muscles attaching to the flexor longus, namely, the flexor accessorius and the 4 lumbrical muscles.

Flexor Digitorum Accessorius Muscle. This muscle lies in the mid-line of the hind sole and originates by 2 heads, one from each side of the calcaneus. Distally, it is attached to the tendon of the flexor digitorum longus. Its function is to hold the long flexor in the midline so as to effect a direct pull on the toes.

Lumbrical Muscles. Four lumbrical muscles arise from the tendons of the flexor digitorum longus. They pass along the medial side of the metacarpophalangeal joints beneath the transverse metacarpal ligament and are inserted into the base of the proximal phalanx and the dorsal expansion. They act to adduct the lateral 4 toes toward the large toe.

The third layer of muscles includes the adductor and the short flexor of the big toe and the short flexor of the little toe.

Flexor Hallucis Brevis Muscle. The short flexor of the big toe is narrow and tendinous at its origin at the cuboid but forms 2 muscular bellies distally, each of which inserts by a tendon into the base of the proximal phalanx. The medial tendon fuses with that of the abductor hallucis, while the lateral tendon inserts with that of the abductor. This

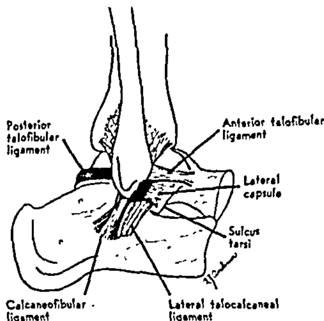


FIG. 445. Schematic drawing showing

subluxation of the ankle joint, J. Bone & Joint Surg. 34A:853)

and the fibula. The mortise, or socket, is deepened posteriorly by the transverse tibiofibular ligament. The lower extremities of the tibia and the fibula are held together by the interosseous ligament and the antero-inferior and the postero-inferior tibiofibular ligaments.

Ligaments. The capsule of the ankle joint is composed of various ligaments joined together. The *anterior ligament* is thin, wide and membranous and extends from the distal surface of the tibia to the neck of the talus. The *posterior ligament* is short and thin and extends from the posterior border of the tibia to the posterior surface of the talus. The strong medial *deltoid ligament* is attached above to the medial malleolus and distally radiates anteriorly to the tuberosity of the navicular, the spring ligament and the neck of the talus; directly below to the sustentaculum tali; and posteriorly to the body of the talus. It assists through the spring ligament in holding up the head of the talus and therefore the longitudinal arch. Three ligaments stabilize the lateral side of the joint. The *anterior talofibular ligament* runs forward from the lateral malleolus to the neck of the talus. The *posterior talofibular ligament* runs backward from the lateral malleolus to the posterior tubercle of the talus. The posterior tubercle may exist as a separate piece of bone attached to the talus by fibrous tissue or cartilage and is known as the os trigonum. The *calcaneofibular ligament* passes downward from the tip of the lateral malleolus to the lateral surface of the calcaneus. When the foot is plantar-flexed, the anterior talofibular ligament lies in a vertical direction and is therefore the ligament most likely ruptured in ankle sprains.

Movements of the Ankle Joint. The socket is wider in front than behind. Similarly, the distal articular surface of the talus is widest. The movements are dorsiflexion and plantar flexion When the ankle joint is dorsiflexed, the talus is wedged tightly between the two malleoli, thus securing stability. In plantar flexion, the narrow posterior part of the talus lies between the malleoli, and a small amount of side-to-side motion is possible. If the intermalleolar interval is narrowed by injury or disease, the anterior talus cannot engage in the mortise, and dorsiflexion is restricted.

Tarsal Joints. The talus forms a gliding synovial joint with the calcaneus. It articulates with a large facet on the middle of the upper surface of the calcaneus and another on the forward portion of the calcaneus for the head of the talus. The strong *interosseous ligament* runs transversely between these 2 facets and holds the 2 bones firmly together. Inversion and eversion movements are possible at this articulation. The distal end of the talus forms a rounded head which articulates in a socket in the navicular. The head of the talus is supported to a large extent by the spring ligament, the *plantar calcaneonavicular ligament,* which with the *lateral calcaneonavicular ligament* binds the calcaneus to the navicular head. The calcaneocuboid joint lies lateral to but in the same transverse plane as the talonavicular joint. These joints, although separate from each other, are described as the *"transverse tarsal joint."* Medial to the cuboid and distal to the navicular lie the 3 cuneiform bones. A synovial-lined cavity exists between each pair of tarsal bones, and these joints frequently communicate with one another.

At all the tarsal joints, the principal movement is gliding in an upward and downward

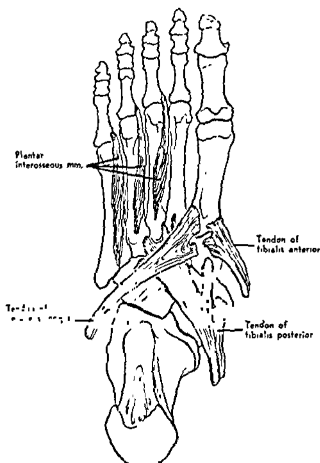


FIG. 444 Deep layer of plantar aspect of foot showing the course and insertions of the invertor and evertor muscles. Note the course of the tibialis posterior which forms a sling beneath the navicular to help form the longitudinal arch. The plantar adducting interossei are also depicted. (After Wood Jones)

forward 3 plantar metatarsal arteries in these spaces. Each of the latter bifurcates into 2 *plantar digital arteries*—1 for each of 2 adjacent toes. At the outer extremity of the arch, the lateral plantar artery of the little toe originates. The first plantar metatarsal artery springs from the junction of the dorsalis pedis with the plantar arch.

The fourth layer consists of the interossei and the tendons of the peroneus longus and the tibialis posterior.

Interossei Muscles. Three plantar and 4 dorsal interossei lie between the metatarsals and arise from them. The plantar interossei spring from the metatarsal of the toe upon which it acts, each inserting into the medial side of the base of the proximal phalanx and into the dorsal expansion. They adduct the lateral 3 toes toward the 2nd toe. The dorsal interossei spring from the sides of each pair of metatarsals. The interosseus in the first space inserts into the medial side of the base of the first phalanx of the 2nd toe. That in the second space inserts into the lateral side of the base of the proximal phalanx of the 2nd toe. The 3rd and the 4th dorsal interossei insert into the lateral side of the base of the proximal phalanx of the 3rd and the 4th toes, respectively. Each of these muscles also inserts into the dorsal expansion. The dorsal interossei abduct the 2nd, the 3rd and the 4th toes from the mid-line of the 2nd toe. They also assist in flexing the metatarsophalangeal joints and extending the interphalangeal joints.

Tendon of the Tibialis Posterior. After this tendon enters the sole, it divides into a larger *medial* part, which inserts into the tuberosity of the navicular and the lower surface of the medial cuneiform bone; and a lateral part which gives off slips of attachment to all tarsal bones except the talus, and also to the bases of the 2nd, the 3rd and the 4th metatarsal bones. The main tendon lies upon the *plantar calcaneonavicular ligament*. The latter extends from the sustentaculum tali to the navicular and, with the tendon of the tibialis posterior, supports the head of the talus. It is the main structure which supports the longitudinal arch and is known as the "spring ligament." A sesamoid bone may develop within the tendon under the spring ligament.

Tendon of the Peroneus Longus. This tendon turns into the sole at the lateral border of the cuboid and passes to its insertion into the base of the 1st metatarsal and the adjacent cuneiform. It is covered by a fibrous sheath. It may develop a sesamoid bone where it enters the groove on the under surface of the cuboid. In addition to its function of everting the foot, the peroneus longus acts as an antagonist to the anterior tibial and stabilizes the first metatarsal for the flexion pushoff of the big toe. Loss of the peroneus longus results in the first metatarsal's assuming a dorsiflexed position and loss of strength of the pushoff.

ANKLE JOINT

The ankle is a hinge joint in which the talus fits into a mortise formed by the tibia

Forcible inversion of the plantar-flexed foot most commonly ruptures the anterior talofibular ligament. With the foot in equinus, this ligament lies vertical in line with the long axis of the fibula. As the force continues, the calcaneofibular ligament is also torn. The posterior talofibular ligament is rarely injured and then only with severe injuries causing complete dislocation.

When the anterior tibiofibular is interrupted, the ankle is relatively stable at 90°; but with the foot in equinus, there is instability in the long axis as well as the vertical axis of the talus. As the foot is plantar-flexed, the talus displaces forward in the ankle mortise a few degrees. The body of the talus can also be tilted a few degrees medially with an inversion strain so that the lateral joint space is slightly widened. When the calcaneofibular ligament is torn, in addition to the anterior talofibular ligament, the medial tilting is greater in degree but has no effect on forward subluxation. If, as rarely occurs, all 3 ligaments are torn, the ankle is completely unstable.

Subluxation of the ankle is often associated with damage to the superior articular surface of the talus, involving mainly the cartilage and simulating osteochondritis dissecans.

The medial collateral ligament, or the deltoid ligament, is composed of very strong fibers. Its component fibers include these ligaments: (1) tibionavicular, (2) anterior talotibial, (3) calcaneotibial and (4) posterior talotibial It is rarely ruptured. Instead, by an abduction force, the internal malleolus is avulsed. As the force continues laterally, the tibiofibular ligaments tear, mainly in their anterior aspect, the rupture extending upward into the interosseous membrane.

Clinical Picture

A history is often obtained of a forced inversion strain with the foot plantar-flexed. Within a few hours, the anterolateral aspect of the ankle becomes greatly swollen, painful and exquisitely tender. Walking causes severe discomfort and instability. Tenderness is well localized to the site of the talofibular and perhaps also the calcaneofibular ligaments. Within *a few days* an area of ecchymosis develops, the blood seeping beneath the deep fascia as far as the toes. Pain can be accentuated by forcefully inverting and plantar-flexing the foot.

If the rupture is mild, symptoms subside within a few days, and normal function is regained. On the other hand, if complete dissolution of fibers has occurred, the acute symptoms gradually subside over a period of weeks, but some tenderness and swelling persist about the lateral aspect of the ankle. This generally follows inadequate immobilization for an insufficient length of time. The ankle is subject to recurrent sprains, and the patient complains of weakness, instability, a giving-way, inability to run, and difficulty in standing on the ball of the foot. Pain is felt when walking on uneven ground. Vague tenderness is perceived in front of the fibula as far as the neck of the talus. Forced inversion strain accentuates the pain.

Roentgenologic Findings

At the initial acute stage, a local anesthetic is injected about the outer side of the ankle. While forcefully plantar-flexing the foot, a lateral view is taken. Forward displacement of the talus from the ankle mortise attests to disruption of the talofibular and possibly also the calcaneofibular ligaments. The anterior view is taken while the foot is forcefully inverted with the foot in equinus. Slight tilting of the talus indicates a tear of the talofibular ligament, while marked tilting implicates both talofibular and calcaneofibular ligaments. Failure to find a defect in the superior articular surface of the talus does not rule out an injury at this site.

Treatment

When no instability is demonstrable by roentgenograms, an acute ankle sprain is immobilized in a plaster cast for a period of 3 weeks If stress roentgenograms reveal displacement of the talus from the ankle mortise, regardless of whether complaints are recent or of long standing, repair of the ligaments should be attempted. In the event that the ligaments have retracted or disintegrated, the Watson-Jones method of tenodesis should be done

Technic.[3] An incision is made posterior and inferior to the lateral malleolus and is carried proximally along the outer aspect of the leg

[3] Watson-Jones, W.: Fractures and Joint Injuries, ed 3, vol. 2, Baltimore, Williams & Wilkins, 1943.



Fig. 446 Depicting instability of the *talus on its long axis (left)* and in the coronal plane demonstrable only on inward and downward strain respectively. (Cases of Drs. K J. Anderson and J. F. LeCocq)

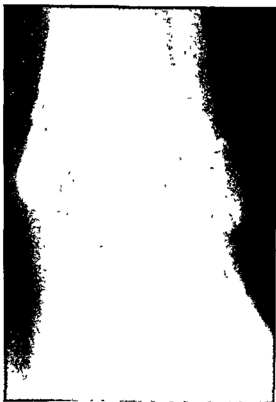


Fig. 447. Secondary ossification center in epiphysis of fibula This attachment of ligaments forms a weak link subject to frequent strains and causes the child to fall.

direction to produce plantar flexion and dorsiflexion of the distal part of the foot. The talonavicular and the calcaneocuboid joints also have *side-to-side gliding which contribute* to inversion and eversion motion. However, this latter movement takes place chiefly at the talocalcaneal joint. In stabilizing the hind foot, it is necessary to arthrodese the talocalcaneal, the talonavicular and the calcaneocuboid joints. This is known as the "triple arthrodesis."

ANKLE SPRAIN

A sprain is defined as a partial or complete rupture of the fibers of a ligament. Any ligament about the ankle may be torn by a force which exerts traction in the direction of the fibers. When the ligament is strong, a fragment of bone is avulsed from the point of insertion of the ligament.

Pathologic Anatomy[1, 2]

The lateral collateral ligament has 3 components:

1. **Anterior Talofibular Ligament.** This is the thinnest and most fragile portion, about 2.5 mm. in thickness, and uniting the anterior border of the fibula to the neck of the talus It is visualized only after removing the overlying capsular tissue.

2. **Calcaneofibular Ligament.** This is *stronger and thicker and* extends from the tip of the fibula to a colliculus on the lateral surface of the calcaneus. It lies extracapsular.

The lateral talocalcaneal ligament occupies the space between these two fasciculi, running parallel with the calcaneofibular ligament and blending not only with the latter *but also with the anterior* talofibular ligament The lateral capsular tissue in the triangular area between the anterior talofibular and talocalcaneal ligaments blends with these structures before passing beneath the calcaneofibular ligament.

3. **Posterior Talofibular Ligament.** This is a very strong band of fibers extending from the fibula to the lateral tubercle of the posterior aspect of the talus

[1] Anderson, K. J, LeCocq, J F, and LeCocq, E A · Recurrent anterior subluxation of the ankle joint A report of two cases and an experimental study J. Bone & Joint Surg 34A 853, 1952

[2] Leonard, M. H : Injuries of the lateral ligaments of the ankle, J. Bone & Joint Surg 31A·373, 1949.

Treatment

Relief of pain is obtained by conservative or surgical means.

Conservative. Rest and abstinence from weight-bearing, application of heat, and salicylates will effectively but temporarily relieve symptoms in most cases. Injection of Meticortelone® is done at weekly intervals. Then, as symptoms are controlled, intervals between intra-articular injections are increased gradually until the maximum interval can be determined.

Surgical. Persistence of symptoms, enough to constitute a continuous disability, is the indication for surgery.

1. Denervation of the Ankle Joint. When preservation of motion is desirable, the distal branches of the tibial and the deep peroneal nerves will eliminate pain effectively. The motor loss (extensor digitorum brevis) is negligible. A neuropathic joint will not develop.[4]

2. Arthrodesis. Elimination of motion permanently relieves pain and provides stability. (These procedures are discussed in the section on poliomyelitis.)

TUBERCULOSIS OF THE ANKLE AND THE FOOT[5]

Pathology

In children the osseous focus in the tibia and the fibula is metaphyseal, the epiphyseal plate constituting a barrier to spread toward the ankle joint, and the infection remains extra-articular. In contrast, the adult lesions are epiphyseal, and spread to the synovium is inevitable. A focus in the astragalus invariably infects the ankle joint above, the subastragalar joint below and the midtarsal joints distally. An os calcis focus, particularly in the posterior half of the bone, can heal without infecting a joint. When a small tarsal bone is infected, it spreads rapidly throughout the entire tarsal area, with osteoporosis and destruction of all small bones. The intercommunicating synovial cavities favor this rapid spread. Metatarsal bones are rarely affected. Their involvement is commoner in infancy. A periosteal reaction similar to spina ventosa is seen. The lesions are usually granular osseous foci occurring in the tibia, the fibula, the astragalus and the os calcis most commonly. The severely acute exudative diffuse infections and the large caseous lesions are uncommon. Abscesses and fistula formation are frequent because of the superficial location. An abscess of the ankle joint generally erupts at a weak spot anterolaterally. Posterior foci in the os calcis point at the sides of insertion of the Achilles tendon. Astragalar abscesses spread over the dorsum of the foot. Subastragalar infections produce an accumulation of fluid on the outer side about the peroneal tendons.

Various deformities result from muscular and capsular contractures. Tibio-astragalar arthritis causes equinus and equinovarus. Subastragalar lesions cause calcaneal dorsiflexion, talipes calcaneus in which atrophy of the calf muscles is associated with contracture of the extensor digitorum communis. Lesions of the scaphoid and the inner cuneiform cause adduction of the forefoot, while those of the anterior os calcis and cuboid cause abduction.

Clinical Picture

A doughy swelling with little or no pain or tenderness and mild increase in cutaneous temperature gradually appears. It tends to subside with rest and to reappear as activity is resumed. Eventually, the swelling is persistent, and pain and muscle spasm are worse. When the tibio-astragalar joint is involved, ankle flexion and extension are limited. Subastragalar lesions cause limitation of eversion and inversion. Passive side-to-side movements of the os calcis are quite painful. When infection involves the entire tarsal area, the entire foot, particularly the dorsum, is markedly swollen, boggy, generally warm and tender; movement in any direction is very limited and painful. An abscess or a fistula may be evident.

Roentgenologic Findings

When the infection is metaphyseal in the tibia, the findings of localized bone destruction and osteoporosis extend only to the epiphyseal line. When infection invades the ankle joint, diffuse osteoporosis and loss

[4] Casagrande, P. A., Austin, B. P., and Indeck, W.: Denervation of the ankle joint, J. Bone & Joint Surg. 33A:723, 1951.

[5] Sanchis-Olmos, V.: Skeletal Tuberculosis, Baltimore, Williams & Wilkins, 1948.



Fig. 448. Brittain arthrodesis of the ankle.

and distally along the lateral border of the foot. The peroneus brevis tendon is severed at its muscular origin, and the muscle is sutured to the peroneus longus. The tendon is passed through a drill hole in the fibula, then through a second drill hole in the neck of the talus, and finally through a third drill hole in the fibula. Then the tendon end is pulled up taut, while the foot is held in forced eversion, and sutured to the distal end of the same tendon. Postoperatively, immobilization is maintained for 6 weeks. Occasionally, slight loss of inversion movement is observed.

DEGENERATIVE ARTHRITIS OF THE ANKLE

Degenerative arthritis of the ankle is due to trauma, acute or chronic infection, rheumatoid arthritis and metabolic disease, especially gout. These processes in effect soften the articular cartilage, which succumbs to the pressures of weight-bearing and the degeneration of advancing age.

Trauma may be of several types. A severe sprain, subluxation or dislocation may appear to be innocent by virtue of a negative roentgenogram, but flakes of cartilage may be intra-articular where they jam and erode the articular surfaces. Torn fibular collateral ligaments, which can be demonstrated by forcefully inverting the hindfoot and taking an A-P view of the ankle, cause instability of the joint, particularly with the foot in equinus Consequently, the articular surfaces are exposed to unequal impacts with production of degenerative changes. Fractures which extend through the joint surfaces may damage the apposing cartilage. When fractures of the malleoli are reduced inadequately, an enlarged mortise will produce unnatural rocking. When a malleolar fracture is reduced and healed while the astragalus is in equinus, the wider anterior portion of the astragalar body will not engage in the mortise, and the foot will remain in permanent equinus. In walking, the astragalus is forced upward against the narrow unyielding bimalleolar interval with resultant damage to the articular surfaces.

Clinical Picture

An antecedent history of trauma, infection, gout or rheumatoid arthritis is often obtained Gradually, symptoms typical of degenerative arthritis appear, namely, pain and stiffness [illegible]

[illegible] capsular tissues. Moderate tenderness is generalized, and joint motion is often restricted

Roentgenologic Findings

The joint space is narrowed, and the subchondral cortices are dense, irregular and often cystic in appearance. Bony projections develop at the margins, particularly at the anterior tibial border. There may be evidence of an old malunited fracture, a punched-out gouty tophus, or an infectious or rheumatoid process.

graft 1 by 2 inches is removed from the tibia immediately above the joint level. This graft is slid across the joint and firmly fixed in a gutter formed by removal of bone from the anterior aspect of the astragalus.

2. *Intramedullary Sliding Cortical Graft.* A tibial cortical graft measuring 1 by 3 inches is removed from the shaft higher up. The distal 1½ inches of tibia is left intact as a collar By a suitable broaching instrument a slot is cut in the medullary canal extending into the astragalus, which has been lined up properly with the tibia. The bone graft is inserted into the opening left by its removal and driven distalward along the canal across the ankle joint and into the astragalus. The tibia and the astragalus are firmly impacted before closure.

A sliding bone graft is contraindicated in children because of necessity it must traverse the epiphyseal line, thereby interfering with growth.

Extra-articular Arthrodesis. This is described for completeness. Some orthopaedic surgeons still are opposed to the intra-articular method. The technic is as follows: A longitudinal incision is made over the posterior aspect of the ankle joint between the Achilles tendon and the medial malleolus. By blunt dissection the capsule is encountered, and the flexor hallucis longus tendon is retracted medially With an osteotome, flaps of bone are turned downward from the posterior aspect of the tibia and upward from the superior aspect of the os calcis. A massive bony bridge thus overlaps the ankle joint from behind.[6]

Excision of Bones. Theoretically, a tuberculous infection may be confined to one bone, and it should be possible to remove the bone and cure the condition Practically, this is not successful. Extension beyond the bone is frequent, and removal of a bone creates a dead space where the disease may run rampant. The risk is too great to warrant the procedure.

Amputation. Extreme destruction and particularly diffuse tarsal infection with abscesses and draining sinuses should be treated by amputation well proximal to the site of involvement.

[6] Campbell, W. C An operation for the induction of *osseous fusion* at the ankle joint, Am. J. Surg 6 588, 1929

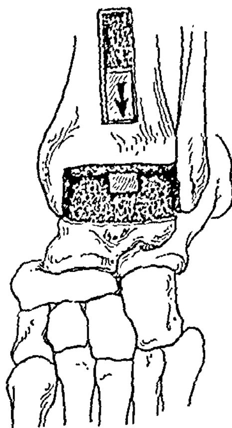


Fig. 449. Intramedullary sliding bone graft for ankle arthrodesis. (Brittain)

STENOSING TENOSYNOVITIS AT THE ANKLE

Where tendons angulate about bony structures on the way to their destination, they are enclosed by a fibrous sheath which acts as a pulley. The sheath may become thickened, usually under conditions of excessive and constant movement of the tendon, and constricts the enclosed tendon. Distal to the point of constriction, the tendon displays a bulbous swelling. Pain is associated with the movements specific for that tendon. Microscopically, the sheath exhibits nonspecific inflammatory and degenerative changes and increase of its fibrous elements. Metaplastic formations of cartilage are seen occasionally.

The condition has been reported involving the tibialis anterior, the tibialis posterior at the ankle, the extensor digitorum longus and the peroneal tendons below the lateral malleolus.[7, 8] The inferior retinaculum, which en-

[7] Hildebrand, O Tendovaginitis Chronica Deformans und Luxation der Peronealsehnen, Deutsche Ztschr Chir. 86 526, 1907.

[8] Burman, M : Stenosing tendovaginitis of the foot and ankle, A.M.A Arch Surg. 67:685, 1953.

definition of the cortex of tibia, fibula and astragalus without narrowing of the joint space are the earliest findings. The joint narrows as the articular cartilage is destroyed. As the infection spreads to the subastragalar, the midtarsal and the distal tarsal joints, osteoporosis, loss of articular margins, narrowing of spaces, and actual collapse of bony structure become evident in these areas.

Prognosis

Extra-articular foci generally heal without articular involvement. An early ankle joint infection may heal without spread to other joints. When the anterior subastragalar joint or other tarsal joint is infected, spread to the remainder of the tarsus is inevitable.

Treatment

Constitutional care includes absolute bed rest, nutritious diet, fresh air and good hygiene. Streptomycin, PAS and INAH are administered A cast is applied from the upper leg to the toes, and the joints are immobilized until they become quiescent. When the lesion is definitely metaphyseal, as in children, immobilization is continued until healing by bony reconstitution is evident. Epiphyseal lesions and infection of a tarsal bone should be regarded as synonymous with joint involvement and spread to other tarsals, although the extent of spread may not be evident clinically or by roentgenograms. Surgical obliteration of joints should be extensive and complete. Otherwise, an overlooked focus may become rampant and so destructive that amputation may be the only indicated treatment. The surgeon should take advantage of the earliest situation to effect a final cure. The only reason for delay is an attempt to improve the general and the local conditions. To reduce the possibility of further destruction and miliary dissemination provoked by surgery, one waits until the hyperallergic state has lessened as indicated by normal temperature, increase of appetite and weight gain, decreased sedimentation rate and, above all, a lymphocyte-monocyte ratio greater than 5. The ankle and the foot are rigidly immobilized until operation is performed.

Intra-articular Arthrodesis. Antibiotics and chemotherapy will not in themselves eradicate a bony focus. The argument against invading the joint and resecting the abnormal tissue because of lighting up the infection and dissemination is no longer valid. The antibiotics and chemical agents form an effective cover under which such surgery may be done.

Technic. An anterolateral incision is made in the lower leg, crossing the ankle over the dorsum of the foot to the base of the third metatarsal. The transverse crural and cruciate crural ligaments are sectioned. The extensor hallucis longus and the anterior tibial tendons with the dorsalis pedis artery, veins and deep peroneal nerve are retracted medially, and the extensor digitorum longus tendons are retracted laterally. The anterior lateral malleolar artery is ligated and cut The anterior capsule may be cut longitudinally, the incision being prolonged upward into the periosteum over the tibia which is exposed by subperiosteal dissection. The articular cartilage is removed from the lower end of the tibia, the lateral malleolus and the superior surface of the astragalus. All abnormal tissue is removed until normal-appearing cancellous bone is exposed. Slivers of bone removed from the tibia are packed into the joint to fill all spaces and to make firm contact. Next, the sinus tarsi is cleared of its fibrofatty tissue The capsules of the subastragalar, the astragaloscaphoid and the calcaneocuboid joints are incised, and articular cartilage is removed down to cancellous bone. Sufficient bone should be removed to correct deformity. The overlying tissues are sutured in layers, and a cast is applied with the ankle in a position of slight equinus and the heel in slight valgus. The cast should extend above the knee in order to avoid rotatory strains at the ankle

Factors Promoting Fusion. Removal of bone and cartilage may be extensive and leave a joint easily distracted and subject to shearing strains, thereby defeating bony bridging of the ankle. The factor of *compression* may be utilized by inserting a pin in the tibia and another in the os calcis and utilizing connecting turnbuckles or the Charnley apparatus to force the bone surfaces together. Slots cut in the cast will permit this without disturbing immobilization Compression may also be obtained by permitting early gentle weight-bearing. *Immobilization by a cortical bone graft* may be added.

1. *Sliding Inlay Bone Graft.* A cortical

The *deep transverse metatarsal ligaments* are 4 strong flat bands which lie between the heads of the metatarsal bones. They are attached to the margins of the plantar ligaments of the capsules. This ligament, in conjunction with the transverse head of the adductor hallucis muscle, prevents the metatarsal heads from spreading apart. The digital nerves and vessels and the lumbrical tendon lie in relation to the plantar aspect of the ligament. Just proximal to this site is the favored location for the development of a neuroma. The interosseous tendons lie on the dorsal surface of the ligament.

Abduction of the large toe is accomplished by the *abductor hallucis* which inserts on the medial aspect of the base of the proximal phalanx. Adduction is performed by the *adductor hallucis*, which inserts with the lateral head of the flexor hallucis brevis into the base of the proximal phalanx. This muscle is composed of two portions. The oblique head arises from the sheath of the peroneus longus and the bases of the 2nd, the 3rd and the 4th metatarsal bones. The transverse head springs from the plantar ligaments of the lateral 4 metatarsophalangeal joints.

METATARSALGIA

Metatarsalgia may be defined as pain about the distal end of the foot at the metatarsophalangeal junctions. In order to interpret the cause of the disability, it is necessary to have a knowledge not only of the anatomy but also of the mechanics of the forefoot.

Mechanics of the Forefoot

In walking, the weight is borne mainly on the 1st metatarsal head, secondarily on the 5th, and to a lesser degree on the 2nd, the 3rd and the 4th. Each metatarsal head thus forms a fixed fulcrum about which the flexor tendon is angulated when the heel is raised off the ground and the toe dorsiflexed before the "take-off." The axis of weight-bearing is situated along the 3rd metatarsal while standing but shifts medially as the heel is raised at the beginning of a step. If the first metatarsal is short or is displaced medially, as in a splay foot, the weight is borne on the 2nd metatarsal head.

The metatarsals are arranged in an arch both in an anteroposterior and in a transverse direction. The transverse arch, in which the central 3 bones lie at a higher level than the peripheral bones, is pronounced proximally at the tarsometatarsal junctions and becomes shallower toward its distal extremity. The term "metatarsal arch" generally refers to the shallow concavity over the plantar aspect of the metatarsal heads. The central 3 heads are elevated by the transverse metacarpal ligaments and the transverse head of the adductor hallucis muscle. This mechanism, although seemingly ineffective during weight-bearing when the arch is obliterated, functions to transfer pressure toward the medial and the lateral metatarsal heads. During the take-off movement of a step, the intrinsic muscles flex the toes and help to elevate the central metatarsal heads off the ground, thus relieving them of pressure. Paralysis of these muscles results in clawed toes, dropped metatarsal heads and the inevitable plantar calluses.

Etiology of Metatarsalgia

The causes of pain about the forefoot are (1) ligamentous, (2) muscular, (3) neuritic, (4) arthritic, (5) traumatic, (6) mechanical, (7) circulatory, (8) metabolic, (9) infectious, (10) osteochondrosis of the metatarsal head and (11) march fracture.

Ligamentous Factors. Stretching of the transverse metacarpal ligaments is due to: (1) *congenital* laxity, which typically results in a flatfoot in which the heel is everted, the longitudinal arch depressed, the metatarsals and the toes widely spread (splay foot) and the forefoot supinated in relation to the hindfoot. (2) *acquired* stretching as a result of overweight, prolonged standing, degenerative changes of old age, and following an acute illness. The central 3 metatarsal heads drop and are prominent in the sole when palpated through the thinned subcutaneous fat. Plantar calluses form, particularly over the 2nd metatarsal head, as a result of abnormally excessive pressure. The underlying adventitious bursa becomes inflamed, tender and painful on weight-bearing.

Treatment. Relief of abnormal pressure against the central 3 metatarsal heads relieves the pain and effects disappearance of the calluses. Nonsurgical methods include (1) placing a felt or rubber pad in the shoe behind the central metatarsal heads; (2) fix-

closes the peroneal tendons, is a favored site.

Stenosing Tenosynovitis of the Common Peroneal Tendon Sheath[9]

This condition occurs in patients over 40 years of age. A history of occupational trauma is often elicited. A palpable, tender thickening is found below the edge of the lateral malleolus. Pain is aggravated by both supination and pronation of the foot. The inferior retinaculum is thickened and constricts the peroneal tendons.

Treatment

In the early stages, immobilization may effect a cure. If pain and swelling are persistent, the peroneal sheath must be excised. In spite of leaving an open sheath, good function without dislocation of the tendons may be expected. If the anterior tibial tendon is involved at the level of the transverse crural ligament, the latter is incised laterally and allowed to heal in the lax position.

THE METATARSOPHALANGEAL REGION

Surgical Anatomy

The metatarsophalangeal articulation is a condyloid joint. It is enclosed in a capsule which is strengthened by thickenings, medially and laterally, by strong rounded *collateral ligaments* and, on the plantar aspect, by a thick, dense fibrous structure, the *plantar ligament,* which is fused with the deep layer of the flexor tendon sheath. The dorsal part of the capsule is very thin and fused with the extensor tendon. The plantar ligament of the first metatarsophalangeal joint is replaced by the sesamoid bones in the 2 tendons of the flexor hallucis brevis.

The tendons of the *extensor digitorum longus* insert into the bases of, and extend, the distal and the middle phalanges of the lateral 4 toes. Each tendon forms an expansion on the dorsum of the toe. The expansion of the 2nd, the 3rd and the 4th toes is joined on the lateral side by a tendon from the *extensor digitorum brevis,* which assists in extending the distal 2 phalanges. In addition, the expansion on its medial and lateral sides receives the interosseous tendons and, on the medial side, the lumbrical tendon which aids extension of the distal 2 phalanges.

The *interossei* have a similar arrangement to that in the hand. They attach to the bases of the proximal phalanges before proceeding distally along the outer and the inner margins of the dorsal aponeurosis. Plantar interossei adduct the lateral 3 toes toward the 2nd toe; dorsal interossei abduct the 2nd, the 3rd and the 4th toes outward.

The *lumbrical* muscles, 4 in number, arise from the tendons of the flexor digitorum longus, cross the medial side of the metatarsophalangeal joints of the lateral 4 toes and insert into the base of the proximal phalanx before joining the dorsal aponeurosis

Flexion of the 4 small toes is accomplished by the flexor digitorum longus and the flexor digitorum brevis. Opposite the 1st phalanx, the short flexor is superficial, then splits into 2 segments to allow passage for the long flexor, which inserts into the base of the distal phalanx. The short flexor is attached to the margins of the middle phalanx.

Therefore, movements of the small toes are as follows: flexion is accomplished by the intrinsic muscles at the proximal joint, the short flexor at the middle joint, and the long flexor at the distal joint. Extension is accomplished by the long and the short extensors, aided by the intrinsics, acting upon the distal 2 phalanges. Adduction is effected by the lumbricals and the plantar interossei, while abduction is done by the dorsal interossei.

In the large toe, the *flexor hallucis longus,* which inserts into the base of the distal phalanx, and the *flexor hallucis brevis,* which inserts into the base of the proximal phalanx, accomplish flexion. On the dorsum, the *extensor hallucis longus* inserts upon the base of the distal phalanx, and the most medial slip of the extensor digitorum brevis attaches to the base of the proximal phalanx. These act to extend the large toe.

The small toe is flexed not only by the flexor digitorum longus but also by a separate muscle, the *flexor digiti minimi brevis,* which arises from the base of the 5th metatarsal bone and inserts into the base of the proximal phalanx. The small toe has only a single extensor from the extensor digitorum longus.

[9] Parvin, R W., and Ford, L T: Stenosing tenosynovitis of the common peroneal tendon sheath, J. Bone & Joint Surg. 38A:1352, 1956.

the metaphysis with the epiphyseal plate, depriving the epiphysis of adequate circulation. The fact that the condition has its onset during the active growth period when the fragile layer of calcified cartilage in the plate is *wide strongly supports* this theory. (See discussion on etiology under "Slipped Femoral Epiphysis.")

Pathology

The epiphysis undergoes aseptic necrosis. Reactive hyperemia and decalcification take place *in the metaphysis*. Capillary inshoots penetrate the cartilaginous epiphyseal plate, absorb and break up the necrotic bone into separate fragments and replace the latter by the process of creeping substitution. Before this happens, the trauma of weight-bearing causes multiple fractures within the necrotic epiphysis and, when the latter is regenerated, it is deformed. Further trauma causes fragments of cartilage to break off and lie loosely within the joint. Degenerative arthritis eventually develops. Spurs form at the margins. The metatarsal shaft develops a compensatory hypertrophy. The second metatarsal is most often affected, less commonly the third, and rarely the others.

Clinical Picture

During the active growth period at puberty, a painful, tender swelling of the soft tissues develops about the affected metatarsophalangeal articulation, usually the 2nd. Weight-bearing and movements of the toe aggravate the discomfort. Symptoms and findings gradually subside within a few weeks. Except for slight, momentary twinges of pain, the foot remains asymptomatic for a number of years The pain-free interval is suddenly interrupted by an incident such as an athletic injury, excessive walking, or a girl wearing her first pair of high-heeled shoes. The metatarsophalangeal joint becomes painful, swollen and tender Pain associated with weight-bearing and movement of the toe is relieved by rest. On examination, the metatarsal head is palpably enlarged, irregular and tender. Often the toe is hyperextended at this joint. With the advent of degenerative arthritis in later years, complaints of pain and stiffness at rest and in humid weather and restricted toe movement are added.



Fig. 450. Osteochondrosis of metatarsal head. (*Top*) Roentgenograms show sclerosis, irregularity, widening, spurring and flattening of the second metatarsal head. (*Bottom*) A frontal view of a resected metatarsal head. The articular cartilage is irregular and dull over most of its surface, and the head is indented. About the margins, particularly at the dorsum, is the thickened capsule containing the bony spurs and infiltrated with cartilaginous tissue. The cleft at the lower margin suggests separation and formation of a loose body.



Roentgenologic Findings

In the early acute stage, the epiphysis appears dense in contrast with the metaphyseal rarefaction. Gradually, it becomes fragmented, *irregular*, widened and flattened at its articular aspect. The periphery of the epiphysis displays spur formation. Early, the articular space is widened. Late, it is narrowed, the opposing bony surfaces being

ing a ¼ inch thickness leather bar, placed transversely, on the sole behind the metatarsal heads; (3) inverting the heel and everting the forefoot by medial and lateral shoe wedges, respectively, or by adhesive strapping (this throws the weight more medially on the 1st metatarsal head); (4) removal of callus by nightly hot soapy soaks followed by the application of a 20 per cent solution of salicylic acid in collodion; (5) strengthening the intrinsic muscles by toe flexor exercises; and (6) weight reduction.

Muscular Factors. Weakness of the intrinsics deprives the toes of strong flexor power, and the metatarsal heads drop. In poliomyelitis, paralysis of the foot dorsiflexors results in an equinus which throws the weight forward on the forefoot. In addition, the common cavus deformity which follows this disease causes a downward tilting of the metatarsals, and added pressure is brought to bear distally.

Treatment. This is discussed in the section on "Poliomyelitis."

Arthritic Factors. Any form of arthritis can affect the metatarsophalangeal joints. In the young and the middle-aged patients, rheumatoid arthritis (q v.) is suspected. Severe degenerative arthritis (q.v.) favors the 1st metatarsophalangeal articulation. Degenerative changes in a single joint other than the first suggests an antecedent osteochondrosis (Freiberg's infraction). The absence of roentgenologic evidence does not rule out early degenerative arthritis in the aged individual. The soft tissues about the joint are swollen, warm and tender to the touch. Passive movement of the toe accentuates the pain

Metabolic Factors. An acute exacerbation of gout (q v.) characteristically develops about the 1st metatarsophalangeal joint. Pain is severe and continuous and aggravated by weight-bearing and movement of the large toe. The soft tissues are markedly swollen, reddened, warm and excruciatingly tender

Treatment. The management of arthritis and gout is discussed in the section on "Affections of Joints."

Traumatic Factors. Prolonged walking will cause a sprain of the transverse metatarsal ligament. Pain occurs throughout the distal metatarsal area on weight-bearing and is intensified by spreading the toes passively Tender spots are localized to the depressions between the metatarsal heads. Some swelling is present throughout the forefoot.

A direct impact, such as by dropping a heavy object on the foot or a fall from a height, will cause a contusion over the metatarsal heads dorsally or on the plantar aspect, as the case may be. Fractures may occur.

A narrow toe box of the shoe will compress the metatarsal heads together and cause an intermetatarsal bursitis.

Treatment. Sprain is relieved by rest, heat, and compressing the metatarsal heads by an encircling elastic bandage. At first, contusion over the metatarsal head is treated by icebags and elevation of the foot to minimize the swelling; later, by warm foot baths A metatarsal pad and the wearing of a sponge-rubber-soled shoe will permit early ambulation.

The toe box of the shoe must be wide enough to accommodate the toes. In addition, the bend in the shoe must correspond to the level of the metatarsophalangeal articulations

Mechanical Factors. Any deformity of the foot which changes the axis of the metatarsal to a more vertical direction throws forward the pressure of weight-bearing. An analogous situation develops when high-heeled shoes are worn, especially when the platform for supporting the heel of the foot lies in an inclined rather than a horizontal plane.

OSTEOCHONDROSIS OF A METATARSAL HEAD

(Freiberg's Disease; Kohler's Disease; Aseptic Necrosis of the Metatarsal Head[10])

Like osteochondrosis of an epiphysis elsewhere, this is characterized by the development of aseptic necrosis of the metatarsal head during the growth period and before the obliteration of the epiphyseal line. As a result of repeated trauma in walking, the epiphysis becomes malformed, and subsequent degenerative arthritis ensues.

Etiology

Many theories have been advanced, including trauma, endocrine disturbance and infection. The pathologic picture suggests that a microfracture has occurred at the junction of

[10] Freiberg, A H . Infraction of the second metatarsal bone, Surg , Gynec & Obst. 19:191, 1914.

sponding 2 toes. The syndrome was first described by Morton in 1876,[11] and the pathology was clarified by Betts in 1940.[12]

Anatomy

The cutaneous branches of the internal and the external plantar nerves emerge in the sole of the foot on each side of the flexor digitorum brevis muscle. They divide into digital nerves which proceed distally between the plantar aponeurosis superficially and the tendons deeply. The internal plantar cutaneous nerve gives off one branch to the medial side of the big toe. It next divides into 3 digital nerves to enter the 3 medial web spaces. Each nerve divides on the plantar aspect of the transverse metatarsal ligament and supplies a branch to the opposing sides of 2 toes. The outermost digital nerve receives a communicating twig from the lateral plantar cutaneous nerve. The latter furnishes the nerve to the 4th cleft and a nerve to the outer side of the little toe. The digital nerve to the 3rd cleft is most often involved in this condition. It lies in relation to the plantar surface of the transverse ligament and therefore cannot be compressed between the metatarsal heads.

The digital arteries arise from the deep plantar arch and run forward between the interossei and the adductor hallucis muscle. Each artery accompanies a digital nerve and lumbrical tendon on the plantar aspect of the transverse ligament, dividing to supply adjacent surfaces of 2 toes. Superficial to these structures are lobules of fat. Dorsal to the ligament lie the interosseous tendons, while still further dorsally between each pair of metatarsal heads lies the large intermetatarsophalangeal bursa.

Pathology[13]

The affected digital nerve, usually the one to the 3 to 4 cleft, displays a fusiform swelling just above the point of division of the nerve. The swelling is asymmetric and often affects the fibers to the 4th toe. Microscopically, the swelling consists of marked connective tissue proliferation which is primarily periarterial. The accompanying digital artery shows periarterial fibrosis, thrombosis and incomplete recanalization. The picture suggests a primary vascular lesion, possibly traumatic, with secondary ischemic nerve changes.

Clinical Picture

The pain is neuralgic, often burning in nature, and limited to the sole beyond the necks of the 3rd and the 4th metatarsal bones and the entire surface of the corresponding toes. Occasionally, it may be confined to the nail bed and the tip of the 4th toe. Other small toes are rarely involved. The pain occurs soon after walking on hard surfaces, especially in high-heeled shoes. It is relieved by rest with the shoe removed. In severe cases, it may persist throughout the night.

Examination

Tenderness is perceived by firm upward and backward pressure in the sole just distal to the 3rd and the 4th metatarsophalangeal joints. Swelling of a tumor is rarely palpable. Hypesthesia is occasionally found on the opposing surfaces of the 3rd and the 4th toes. A resistant, very painful ulcer rarely may develop on the inner surface of a toe. Roentgenograms are negative.

Treatment

The neuroma must be removed. Relief of pain is immediate and permanent.

Technic. A longitudinal incision is made in the third web. When the transverse ligament is cut, the metatarsals may easily be spread apart. The nerve is traced proximally and distally, the neuroma is identified, and the whole nerve with its vascular bundle is removed. Postoperatively, immediate ambulation is permitted. Sensation returns to adjacent sides of the toes within a few weeks.

HALLUX VALGUS (Bunion)

Definition

Hallux valgus is a deformity in which the large toe is deflected laterally, and a bony prominence develops secondarily over the

[11] Morton, T. G.: A peculiar and painful affection about the fourth metatarsophalangeal articulation, Am. J. M. Sc, N S. 71:37, 1876.

[12] Betts, L O: Morton's metatarsalgia, neuritis of fourth digital nerve, M. J. Australia 1:514, 1940.

[13] Nissen, K. I: Plantar digital neuritis, J Bone & Joint Surg. 30B:84, 1948.

irregular, sclerotic and marginally spurred. Thickening of the metatarsal shaft is a late finding. Multiple osseous fragments may lie within the joint space.

Treatment

In the acute early stage, rest and removal of weight-bearing are provided by a cast and crutches, in an effort to minimize deformity. Later, symptoms may be relieved by rest, wearing a metatarsal bar or pad, avoiding a high-heeled shoe, and injection of hydrocortisone into the joint. Persistent symptoms are an indication for removal of the metatarsal head.

MARCH FRACTURE
(Perimetatarsal Osteoma)

The slender shaft of a metatarsal bone, usually the 2nd, rarely the 3rd, may yield to unusual stresses and strains such as a prolonged hike. A short first metatarsal which imposes excessive weight-bearing pressure upon the head of the 2nd metatarsal contributes to this condition. The active, energetic child or adolescent is predisposed. A transverse fracture occurs at the middle of the shaft. The fragments remain undisplaced because of the intact thick periosteal envelope which is peculiar to the young age group. Eventually, absorption at the fragment ends, and subperiosteal callus formation reveals the true nature of the condition.

Clinical Picture

The individual complains of pain in the metatarsal area on weight-bearing The dorsum of the foot is edematous, and tenderness is well localized to the middle of the affected metatarsal. By grasping the metatarsal head and manipulating it forcefully, the pain can be reproduced. After several weeks, the pain subsides, and a hard, fixed, nontender enlargement of the metatarsal is palpable.

Roentgenologic Findings

The x-ray film is negative during the first 2 weeks. Then there develops an ellipitical opacity of callus which encircles the middle of the shaft of the 2nd or the 3rd metatarsal. Beneath this the shaft displays a transverse line of translucency, indicating a fracture. The fragments are well aligned and apposed. Complete union and restoration of the bone takes place in 3 months.

Treatment

Simple immobilization in a cast for 3 weeks is sufficient to relieve the discomfort. Thereafter, ambulation is permitted without jeopardy to eventual healing of the fracture.

PLANTAR DIGITAL NEUROMA
(Plantar Digital Neuritis; Morton's Metatarsalgia)

The development of a neuroma in a digital nerve just proximal to its point of bifurcation is associated with severe neuralgic pain in the distal end of the sole referred to the corre-

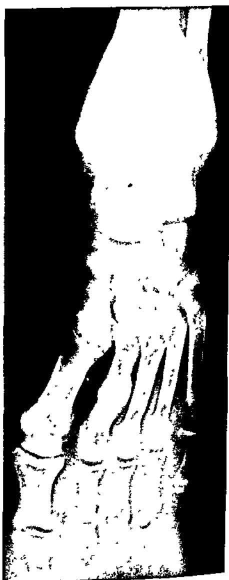


Fig. 451. March fracture.

netatarsals are prominent in the sole, where bnormal pressures result in calluses.

Roentgenologic Findings

These include lateral displacement of the arge toe, degenerative arthritic narrowing [illegible] joint, [illegible] The [illegible] d 1st netatarsal, widening of its base, and abnormal obliquity of the articulation with the 1st cuneiform.

Clinical Picture

The deformity varies in degree of displacement and rigidity. With hallux rigidus, active flexion, which is necessary to take-off in walking, is lost; this is very disabling. The soft tissues over the medial bony prominence are thickened and may be reddened, edematous and tender because of an acute bursitis. The medially disposed 1st metatarsal gives a widened appearance to the foot.

Treatment

Conservative. This is sufficient for mild deformities or where contraindications for surgery exist. Removal of pressure from the medial bony prominence relieves the bursitic pain. A shoe with an enlarged toe portion to

Fig. 453. Hallux valgus (A) Depicting direction of muscle pull which maintains deformity. (B) Shows postoperative condition. The muscle pull is now actively correcting the deformity.



FIG. 452. Hallux valgus. The 1st metatarsal is fixed in abduction (away from the 2nd metatarsal), and the 1st phalanx is subluxated laterally. In addition, the abductor hallucis tendon has become displaced laterally beneath the metatarsal head, as indicated by lateral displacement of the sesamoids, and the muscle is ineffectual in maintaining abduction. Correction requires (1) restoring adduction to the 1st metatarsal by osteotomy or wedge resection and arthrodesis at the metatarsocuneiform joint; (2) transferring the insertion of the abductor hallucis to a more medial and distal position; (3) removal of the medial bony prominence.

medial aspect of the 1st metatarsal head and neck.

Etiology

Heredity. Particularly predisposed is the congenital splay foot in which the metatarsals are widely spread and the 1st metatarsal is shortened and abducted.

Mechanical. Pressure of narrow shoes

Osseous. Wedging of the medial cuneiform causes the 1st metatarsal to angulate medially. Hallux valgus is a secondary effect.

Arthritis. Particularly degenerative arthritis of the 1st metatarsophalangeal joint

Flatfoot. Dropped longitudinal arch causes dorsiflexion at the 1st cuneiform-metatarsal joint; the distal end of the first metatarsal is displaced dorsalward and medially; in consequence, the tensed adductor hallucis draws the big toe laterally.

Pathology

The large toe is displaced lateralward and is rotated about its long axis so that the nail faces medially. The soft-tissue structures on the lateral side, the conjoint tendon of the adductor and the short flexor and the lateral capsule, become adaptively contracted. The sesamoid in this conjoint tendon is enlarged and displaced into the interval between the metatarsal heads. On the medial side, a bony enlargement develops about the metatarsal head and neck. Continued pressure over the bony prominence results in formation of an adventitious bursa. The bursa frequently becomes inflamed and thickened It disappears when the abnormal pressure has been removed. Degenerative arthritic changes frequently develop in the metatarsophalangeal joint, either in cases of long standing as a result of abnormal mechanical strains or as part of a generalized arthritis The joint becomes narrowed and rigid (hallux rigidus) Many cases exhibit the metatarsus primus varus deformity stressed by Lapidus as the main causative factor. This consists of medial angulation of the 1st metatarsal, which is shortened. The deformity arises in consequence of wedging of the first cuneiform or obliquity of the base of the metatarsal so that the latter articulates in a varus position. The base of the metatarsal is widened and develops a bony enlargement which impinges laterally on the base of the 2nd, thereby obstructing replacement.

Associated Pathology. The 2nd toe is displaced dorsalward by the large toe and develops a hammer toe deformity and callus on the dorsum. The foot frequently is flattened, with the heel in marked valgus The transverse arch is flat, and the heads of the middle

skin flap is retracted medially, and a semicircular flap is cut out of the soft tissue over the medial bony prominence. The base of the flap is distal and remains attached to the base of the proximal phalanx. As the flap is elevated, it includes the periosteum over the exostosis. It may also include the adventitious bursa which need not be removed. The exostosis is removed flush with the surface of the shaft, and the cancellous bone which is exposed is covered with bone wax. Toward the plantar aspect, the conjoined tendon of the abductor hallucis and the medial head of the flexor brevis are isolated and severed from their insertion into the plantar portion of the base of the 1st phalanx. They are resutured to the phalanx in a more medial position while the toe is held in the medially corrected position. A drill hole is made in the neck of the metatarsal, and the lateral conjoined tendon with its catgut strands is drawn through and tied in place while the metatarsal is pushed laterally. This aids in overcoming the medial displacement of the metatarsal. Then the medial flap is sutured in a more proximal position. The skin is closed, and a compression dressing aids in securing hemostasis.

Bone Excision Procedures. The rationale is (1) removal of sufficient bone so as to relax the soft-tissue structures and permit correction of the deformity; (2) arthroplasty and mobilization of an osteoarthritic joint. The Keller procedure is the most popular.

Keller Operation. Through a dorsomedial incision, the proximal half of the proximal phalanx is exposed subperiosteally and removed. The redundant periosteum is sewn over the remaining distal half of the phalanx. The exostosis is resected. The position of the toe is overcorrected, and the soft tissue is sutured.

Mayo Operation. The metatarsal head is removed, and a soft-tissue flap is interposed between the metatarsal and the 1st phalanx, constituting an arthroplasty. However, the 1st metatarsal head is an important point of weight-bearing and should be preserved. This can be accomplished by removing only about ¼ inch from the entire articular aspect, preserving the contour of the articulation. The Mayo operation is rarely indicated, as the Keller procedure obtains the same result and preserves the important metatarsal head.

Correction of Metatarsus Varus Deformity. The attack is directed mainly at the metatarsocuneiform joint. The joint is exposed by a dorsomedial incision between the bases of the inner 2 metatarsals. The dorsalis pedis artery is identified and protected. A wedge of cartilage and bone, with the base laterally, is excised from the lateral side of the first metatarsocuneiform joint, permitting correction of the varus. The adjacent medial aspect of the base of the second metacarpal is freshened. Bone chips are packed into the area to promote a fusion. The remainder of the operation consists of the McBride, Keller, or other technic to correct the displacement of the large toe, to excise the exostosis and, if necessary, to interpose a fascial flap. Postoperatively, the foot is immobilized in a cast for 4 weeks.

HALLUX VARUS

In this deformity, the large toe is angulated at the metatarsophalangeal joint toward the medial side. It is usually congenital and associated with other congenital defects. Other causes include: overcorrection of a bunion operation, paralysis of the adductor hallucis, infection about the metatarsophalangeal joint, and malunion of fractures about the joint. This condition must not be confused with metatarsus primus varus in which the angulation occurs in the metatarsocuneiform joint. The clinical description and treatment are included in the section on "Congenital Deformities."

THE LONGITUDINAL ARCH AND FOOT STRAIN

The longitudinal arch of the foot extends from the calcaneus to the metatarsal heads. It is highest medially, the apex being at the navicular, and shallow laterally, where it is limited by the lateral border of the foot coming in contact with the floor. The arch is maintained by (1) the structure of its skeletal components, particularly the relationship between the talus and the calcaneus; (2) the ligamentous support, especially the spring ligament, the interosseous ligament, and the long and the short plantar ligaments; (3) the muscle tone, as resides mainly in the anterior and the posterior tibial muscles. The function of the longitudinal arch is to provide a resili-



FIG. 454. Surgical correction of hallux valgus (1) Capsuloperiosteal flap. (2) Bony prominence removed. (3) Abductor hallucis tendon sutured distally under tension. (4) Optional drill hole for fixation of conjoined adductor-flexor tendon. (5) Sesamoid to be removed.

accommodate the bunion is effective. A thick felt ring placed about the exostosis or a plastic covering cap also relieves pressure. A severely painful bursitis may require puncturing or incising and application of hot moist compresses. In the presence of a suppurative infection of the bursa, incision and drainage followed by warm soaks and administration of antibiotics is indicated When osteoarthritis of the metatarsophalangeal joint is symptomatic, intra-articular injection of hydrocortone, heat, rest and salicylates constitute the treatment.

Surgical. Relief of symptoms is the only indication for surgical correction. It is not undertaken for cosmetic reasons. The type of operation selected is one adapted to the conditions as they exist in the individual case. The following procedures are described as specific for certain situations, but they may be combined and adapted to the case at hand

PLASTIC OPERATION ON SOFT TISSUES (MCBRIDE). A longitudinal incision is made over the dorsolateral aspect of the large toe, extended proximally over the interval between the first 2 metatarsal heads, then crossed over the metatarsal neck medially. The intermetatarsal space is developed deeply to expose the conjoined tendon of the adductor hallucis and the flexor hallucis brevis where it inserts into the medial and the plantar aspects of the first phalanx. It is severed at its attachment, and the sesamoid within its substance is removed A crisscrossed suture of chromic catgut is placed in the tendon, and suture ends emerge from the distal tendon end and are held with forceps. The lateral capsule of the joint is cut. The toe should now be easily moved medially. The

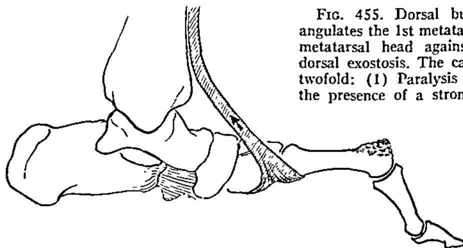


FIG. 455. Dorsal bunion. The anterior tibial angulates the 1st metatarsal dorsad. Pressure of the metatarsal head against the shoe produces the dorsal exostosis. The causes of this deformity are twofold: (1) Paralysis of the peroneus longus in the presence of a strong anterior tibial. (2) Paralysis of the extensors of the big toe in the presence of strong flexors. (See discussion of The Elevated First Metatarsal [Dorsal Bunion], p. 282.)

2. Acquired

A. *Osseous.* Fractures or disease, especially of the talus and the os calcis, may result in a flatfoot deformity.

B. *Ligamentous.* Dislocations cause tearing and eventual lengthening of the plantar ligaments.

C. *Muscle Imbalance.* Weakness of the tibials in the presence of strong peronei causes a valgus deformity. This is discussed more fully in the section on "Poliomyelitis and Cerebral Spastic Paralysis."

D. *Postural or Static.* Internal tibial torsion secondarily causes a flatfoot. In an effort to point the toes forward in walking and standing, the foot is everted. Excessive weight, muscle fatigue, faulty footwear and bad walking habits cause dropping of the longitudinal arch. An antecedent inherent weakness or laxity of the ligaments must be present for a flattening of the arch to occur. This presupposes a congenital hypermobile flatfoot of a mild degree, the deformity taking place on superimposition of postural or static factors.

E. *Arthritic.* Arthritis in the tarsal joints, particularly of the rheumatoid variety, causes reflex peroneal spasm with consequent valgus deformity. If the arthritis and the spasm persist, the flatfoot becomes rigid and permanent. Other types of flatfoot by interfering with the mechanics of tarsal motion will eventually develop degenerative arthritis of the tarsal joints and a rigid foot.

HYPERMOBILE FLATFOOT

This is a congenital type of flatfoot with a marked hereditary tendency displaying extreme mobility of the tarsal and the subtalar joints, a deformity which disappears when the feet are freed of weight-bearing and a short tendo achilles, which limits dorsiflexion at the ankle joint. The condition very probably results from an unstable architecture of the tarsal bones.[14] Other theories include a congenitally short tendo achilles,[15] and weakened muscle power,[16] the deformity being secondary.

Clinical Picture

History. The deformity becomes manifest in childhood. Other members of the same family may be affected. Soon or late, but often in adolescence, pain and fatigue occur on walking or standing. The child has a limited capacity for indulging in sports or other heavy activities. The disability becomes continuous and progressively worse.

Flat Feet Only on Weight-Bearing. When weight is removed, the feet assume the normal shape. In standing, the short tendo achilles is compensated by excessive mobility of the feet, which permits placing the foot flat upon the floor by creating the deformity. The patient can correct the deformity voluntarily by standing on tiptoe or by contracting the long toe flexors and the tibials.

Shortness of Tendo Achilles and Limitation of Dorsiflexion at the Ankle Joint. Instead of the normal 10° to 20° of dorsiflexion beyond a right angle, motion is limited to less than the right angle. To demonstrate this, the forefoot is grasped and forcefully inverted to eliminate the possibility of dorsiflexion at the midtarsal joints. Then the foot is pushed upward, and the angle which the foot forms with the leg is noted.

Hypermobility of Subtalar and Midtarsal Joints. Because of undue ligamentous laxity, the forepart of the foot can be bent passively outward and upward to an unusual degree. This motion is associated with valgus of the heel, bulging of the inner margin of the foot, and depression of the head of the talus. The displacement of these structures permits the heel to come down on the ground in spite of the short tendo achilles

The Deformity. This varies in severity. The forepart of the foot is swung out, forming an obtuse angle with the hind part, the apex being medial. The foot is rotated externally in relation to the leg The head of the talus points downward and inward, forming a prominence on the medial aspect of the foot. The calcaneus is tilted into valgus. The whole sole of the foot is in contact with the ground.

[14] Harris, R I, and Beath, T Hypermobile flat foot with short tendo achilles, J Bone & Joint Surg 30A 116, 1948

[15] Gocht, H: Sehnenoperation beim Pes planovalgus, Ztschr. Orth Chir 14:693, 1905.

[16] Hoke, M.: An operation for correction of extremely relaxed flat feet, J. Bone & Joint Surg. 13:773, 1931.

ent spring for weight-bearing and forward propulsion in walking. The longitudinal arch is generally considered to be absent at birth but gradually appears with development of muscle tone. The thick fat pad in the sole of the infant's foot causes an exaggerated flatfoot appearance.

Normally, with movement at the intertarsal joints, the play between the bones is free, and stability is attained chiefly by muscle power. At the extremes of a movement, the ligaments become taut and help to restrain further movement. When the muscles are fatigued by excessive activity, overweight, or illness, the ligaments are severely stretched, and pain is caused. If the calcaneus lies directly beneath and supports the talus, stability is provided, the ligaments are not strained excessively, and pain is less likely When the foot is quite supple and the ligaments are lax, displacement of the tarsals can occur to an extreme degree before the ligaments can be strained. Such a foot is less likely to incur an acute strain. An acute strain occurs most commonly in a rigid foot, especially when affected by arthritis.

Clinical Picture

The patient complains of pain over the inner border of the foot with standing and walking; this pain is relieved by rest. Tenderness is perceived over the strained ligament. Often the area is localized at the under surface of the navicular or at the anterior and the posterior extremities of the arch. Passive dorsiflexion of the foot intensifies the discomfort. Plantar flexion is painless. Injection of a local anesthetic into the affected ligament relieves the pain.

Treatment

Rest and application of heat relieve the acute pain Repeated needling of the ligament with a local anesthetic expedites recovery. The tension on the ligaments is alleviated by wearing a flexible arch support, adhesive strapping, reduction of weight, and limiting activity. Muscle-building exercises are directed toward strengthening the invertors and the plantar flexors. The arch supports are discarded as soon as possible, because they prevent the stretch reflex which is necessary to maintaining muscle tone. Repeated manipulations, under local anesthesia if necessary, are required occasionally for theoretical elongation of the ligaments and for reduction of the tendency to strain. Various physical therapy procedures include massage, contrast foot baths and faradic stimulation of the small muscles of the foot.

✓Flatfoot

A flatfoot is essentially one with depression or complete loss of the longitudinal arch. As a result, the bony structure, the ligaments and the muscles are altered, and the typical planus or planovalgus deformity ensues. The forefoot is in abduction and slight supination the navicular and the head of the talus are prominent on the medial aspect of the foot the calcaneus is everted (valgus), and the longitudinal arch is depressed. The head of the talus, instead of being supported by the anterior end of the os calcis is displaced medially and downward so that the entire talus angulates forward, downward and medially The entire os calcis is also angulated medially its posterior extremity being displaced laterally and the anterior extremity medially. Secondary changes take place in the bones and the soft tissues when the deformity has existed for some time. The navicular, the cuneiforms and the cuboid become wedge-shaped, the apex of each being directed laterally and dorsally. The plantar, the calcaneonavicular (spring) and the deltoid ligaments are stretched, while the dorsal and the lateral ligaments are contracted. The anterior and the posterior tibial tendons and the plantar muscles are stretched, while the Achilles tendon becomes shortened.

Types of Flatfoot

A flatfoot is congenital or acquired.

1. Congenital \a\57

A. *Hypermobile Flatfoot*. This is a hereditary type, exhibiting marked laxity of ligaments. The midtarsal and the subtalar joints are hypermobile, and the tendo achilles is short.

B. *Rigid Flatfoot with Tarsal Anomalies*. This very common type is due to a bridge, consisting of bone, cartilage, or fibrous tissue, between the talus and the os calcis or between the navicular and the os calcis.

6. Flattening of the longitudinal arch in a lateral view

7. Downward pointing of the talus seen in a lateral view. Normally, it forms a straight line with the navicular, the inner cuneiform and the 1st metatarsal.

Treatment

The treatment, whether conservative or surgical, depends upon the degree of disability. Conservative methods should be tried in all cases. Incapacitating symptoms which resist simpler modes of treatment justify surgical intervention which is aimed at correction of the deformity and arthrodesis of the subastragalar joint. Unless disability in childhood is extreme, arthrodesing procedures should await full bone growth. Before the age of 10, surgical obliteration of tarsal joints is difficult to obtain because of the excessive cartilaginous component of tarsal structures. Also, subsequent bony growth is retarded. If at all possible, surgery should be delayed until the age of 10 and preferably 15.

When the child is severely disabled, the Grice procedure is available. This is an extra-articular arthrodesis by the use of bone struts placed lateral to the talocalcaneal joint. It does not interfere with bony growth and therefore can be utilized during the growth period.[17]

Symptoms generally increase in severity with advancing age. Increased body weight, diminishing muscle strength, increased activity, and the development of osteoarthritis all contribute to the progressive disability.

Conservative Treatment. The aim is (1) to relieve ligamentous tension, (2) to transfer the body weight to the outer side of the foot and (3) to strengthen the invertors and the plantar flexors with which the bones and the ligaments maintain a reciprocal relationship.

1. Arch Supports. Felt pads or sponge rubber inserts glued into the shoe provide a resilient support for the longitudinal arch. When pain and disability are severe, a firm appliance, such as the Whitman valgus brace, made of aluminum, grasps and holds the heel in inversion. It must be individually made from a cast of the foot. It consists of a broad plate which extends over the sole from the middle of the heel to a point behind the 1st metatarsal head. It is wide and acutely curved upward on the medial side to conform with the normal longitudinal arch. Laterally, it is narrow and curved upward to embrace the calcaneocuboid joint. The degree of arching of the plate should be increased at intervals.

2. Shoes. The counter must be firm enough to grasp the heel The waist must be well fitted and tight over the instep to restrict dorsiflexion at the midtarsal joints. At the toebox, adequate room is permitted for plantar flexion of the toes. The heel must be low enough to gain the benefit of heel-cord stretching. A Thomas heel or a ¼ inch wedge on the inner border of the heel throws the weight to the outer border. The shank is rigid or semirigid. The forepart of the shoe has an inward flaring to favor adduction.

Shoes and supports are a measure to reduce

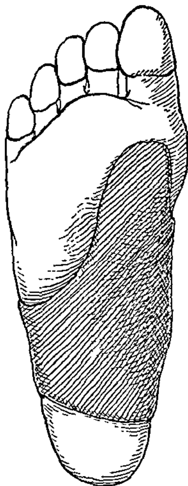


Fig. 457. Whitman arch support.

[17] Grice, D S. An extra-articular arthrodesis of the subastragalar joint for correction of paralytic flat feet in children, J. Bone & Joint Surg. 34A:927, 1952.



FIG. 456. Flatfoot, lateral view in the weight-bearing position. The longitudinal arch is depressed; the astragalus points directly downward instead of forward in line with the scaphoid and the metatarsals.

In long-standing deformity, calluses develop over the medial bony prominences, and the peronei are adaptively shortened.

SYMPTOMS

Symptoms are typical of disability associated with any type of flatfoot. At first the feet feel uncomfortable, burning sensations are experienced, and easy fatigue is noted. The gait becomes clumsy. Pain is more severe when standing than when walking or running. It is due to ligamentous strain and often is felt about the navicular, the talocalcaneal joint, below the medial malleolus, or at the anterior or the posterior extremities of the plantar ligaments. These sites are tender. Edema of the dorsum of the foot is common. In walking, the patient avoids raising the heel before the ball of the foot to prevent strain upon the plantar ligaments. Instead, he raises the entire foot at one time.

PATHOLOGY

The head of the talus lies far medial to the center of the anterior end of the calcaneus. The sustentaculum tali is narrow and supports only the neck of the talus. With superimposed weight, the lack of anterior support causes the talus to be displaced further medially and thrust downward, the calcaneus secondarily being tilted into valgus. The abnormal laxity in the subtalar and the midtarsal joints deprives the tendo achilles of tension stresses which are necessary to promote elongation of the tendon. In consequence, the tendo achilles remains short. To effect dorsiflexion of the forepart of the foot, it is forced into abduction, and the plantar structures are stretched. The longitudinal arch is flattened. All ligaments on the medial and the plantar aspects of the tarsal area are stretched. The tibials and the plantar muscles become elongated and weakened. The peronei become adaptively shortened. Eventually, the tarsal bones assume a configuration adapted to their altered position. They become asymmetrically wedge-shaped, their apices being directed upward and outward. The altered mechanics of tarsal motion lead to traumatic degenerative arthritis and secondary deformities such as hallux valgus.

ROENTGENOLOGIC FINDINGS

Supero-inferior, lateral and oblique projections are taken.

Characteristic findings are:

1. Medial displacement of the head of the talus
2. Anterior projection of the head of the talus
3. A short, tonguelike sustentaculum which does not extend sufficiently far forward to support the head
4. Divergence of the long axis of the talus from that of the calcaneus
5. Excessive amount of head and neck of talus not superimposed on the underlying calcaneus

supination and the tendon pulled distally while it is sutured. Finally, the flaps are sutured together, and the wound is closed. Postoperatively, the foot is immobilized for 6 weeks, followed by a shoe with a thick inner sole and a heel wedge.

TARSAL COALITION
(Congenital Rigid Flatfoot; Peroneal Spastic Flatfoot)

The tarsals originate from mesenchyme and differentiate into cartilaginous precursors of adult bones separated by joints. Ossification is progressive throughout childhood and is completed at puberty. Incomplete division may leave a connecting bar between the talus and the calcaneus and between the navicular and the calcaneus. Varying degrees of interruption in continuity between the two involved bones is represented by a thin or a thick bar composed entirely of bone (synostosis), or bone cleaved in two parts by cartilage (synchondrosis) or fibrous tissue (syndesmosis). The accessory bone, the os calcaneum secundarium, seen between the anterior extremity of the calcaneus and the navicular, probably represents incomplete ossification of the bridge. The talocalcaneal bar is found about the sustentaculum on the medial side of the foot. The effect of the tarsal coalition is to hold the calcaneus in an everted position, the peronei being adaptively shortened. These muscles are not in spasm, so that the term "peroneal spastic flatfoot" is a misnomer Spasm of the peronei rarely occurs reflexly from an irritative lesion within the tarsus, such as a rheumatoid arthritis or an acute strain. Therefore, a rigid flatfoot occurring in childhood is most commonly due to tarsal coalition.

Pathology[20, 21, 22]

The most common type is a bridge of bone springing from the medial surface of the talus, spanning the subtalar joint, and meeting a mass of bone from the medial surface of the calcaneus at the posterior end of the sustentaculum tali (synostosis talocalcanea). The bridge may be interrupted by an accessory joint (articulatio talocalcanea accessoria), a fibrous junction (syndesmosis coalescentia talocalcanea), or a cartilaginous junction (synchondrosis coalescentia talocalcanea). An accessory tarsal bone, the os sustentaculi, occasionally found at the posterior end of the sustentaculum tali, probably represents an incomplete bridging. The all-inclusive term for this condition is *"talocalcaneal bridge."* The firm fixation of the talus to the calcaneus interferes with normal motion at the talonavicular joint. As a result, a degenerative arthritis of this joint is manifest by a characteristic lipping which develops on the superolateral margin of the head of the talus. The

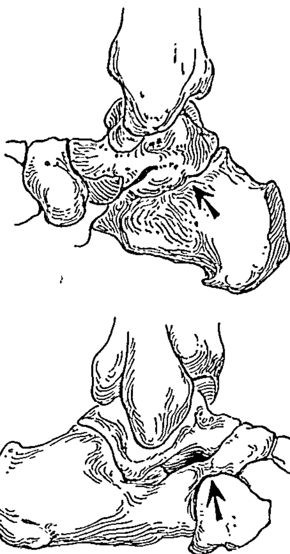


Fig. 458. Tarsal coalition. (*Top*) Talocalcaneal bridge. (*Bottom*) Calcaneonavicular bridge.

[20] Harris, R I, and Beath, T Etiology of peroneal spastic flat foot, J. Bone & Joint Surg. 30B·624, 1948.

[21] Sloman, N On coalitio-calcaneonavicularis, J Orthop Surg. 3 586, 1921.

[22] Badgley, C. E · Coalition of the calcaneus and the navicular, Orthop Surg 15 75, 1927.

the load ordinarily borne by muscles. They do not alter the underlying structural fault nor do they encourage in any way redevelopment of the arch.

3. EXERCISES are designed to improve strength in the invertors and the plantar flexors. A mild or moderate disability may be improved by restoring the balance of muscle power. At the same time, the tendo achilles and the peroneal muscles are stretched repeatedly.

A. *Non-weight-bearing Exercises.* These should be performed in a hot tub bath. The toes are made to plantar-flex and curl over a wash cloth until the patient is able to pick up the object with his toes. The toes may be made more flexible by passively manipulating each toe into flexion The movement is often accompanied by a loud snapping noise and is *followed immediately by freer motion in a* plantarward direction. Next, the foot is actively adducted and, at the same time, dorsiflexed in a medial direction. If the movement is performed properly, the anterior tibial tendon should stand out prominently.

B. *Weight-bearing Exercises.* The patient is instructed in heel-to-toe walking, the toes pointing forward, and weight is borne on the outer border of the foot. A *supination board* is useful. This consists of two boards which are joined at an angle of 135° to 165°. The patient walks forward and backward on the inclined surfaces. Walking or playing in soft sand necessitates strong plantar flexion of the toes and provides excellent exercise for children Ballet dancing should be encouraged.

C. *Specific Resistance Exercises.* The motions of toe flexion, adduction, inversion, and inversion plus dorsiflexion are performed against resistance These must be done slowly, the opposition offered to each movement being light at first and gradually increased at each session. One must be careful not to overexert and strain the contracting muscle

Surgical Treatment. For severe types of deformity and disability, supports, shoes and muscle retraining offer only palliative relief, and the semi-invalidism requires more energetic measures. Various procedures have been devised, some supposedly for merely elevating the longitudinal arch, but all gain their effect by restricting subtalar and midtarsal motion and providing stability for the head of the talus. Arthrodesing operations should not be undertaken in childhood, as the excessive cartilaginous composition of the tarsals makes for difficulty in obtaining a fusion, and bony development may be compromised. The following are commonly employed procedures The patient must be prepared to accept *permanent loss of inversion-eversion motion* Achilles tendon lengthening may be performed at the same time or as a preliminary procedure.[18]

TRIPLE ARTHRODESIS. The technic is described in the section on "Poliomyelitis" When wedges of bone are removed, their bases should be directed downward and medialward, the extent of removal depending upon the amount of correction desired The valgus of the os calcis is corrected so that the anterior end of the bone lies beneath and supports the head of the talus A small amount of heel valgus is normal. The forefoot is pronated at the midtarsal joints. Arthrodesis must not be performed before the age of 10 and preferably at about 15.

When an accessory scaphoid (prehallux) appears as a supernumerary bone on the medial surface of the scaphoid, the posterior tibial tendon is usually attached to it rather than passing beneath and supporting the scaphoid. The loss of the longitudinal arch can be restored by resecting the accessory scaphoid (which prevents adduction of the foot by impinging on the medial malleolus) and transplanting the tendon.

Technic.[19] A longitudinal incision is made from the internal malleolus to the base of the first metatarsal bone. The fascia and the periosteum are incised longitudinally and are reflected inferiorly from the scaphoid and the head of the talus. The posterior tibial tendon is freed from all its attachments except that to the inferior aspect of the medial cuneiform Then it is transplanted into a groove cut on the inferior surface of the scaphoid and held in place by suturing it to the inferior flap The foot should be held in adduction and

[18] Crego, C H, and Ford, L. T.: An end result study of various operative procedures for correcting flat feet in children, J Bone & Joint Surg 34A:183, 1952

[19] Kidner, F. C. The prehallux (accessory scaphoid) in its relation to flat foot, J. Bone & Joint Surg. 11 831, 1929.

Roentgenologic Findings. The talocalcaneal bridge is demonstrated by an axial view of the calcaneus. The x-ray tube behind the heel is directed downward and forward at an angle of 45°. By bending the knees and dorsiflexing the foot at the ankle, interference by soft-tissue shadows is eliminated. When a talocalcaneal bridge is present, the medial side of the subastragalar joint is obliterated. A secondary change seen in the lateral view is marginal lipping on the dorsal aspect of the talonavicular joint.

The calcaneonavicular bar is visualized by an oblique projection of the hind foot. The foot is placed in the prone position, the dorsolateral surface of the foot lying against the casette. The x-ray beam is directed perpendicular to the table.

Treatment

Acute symptoms are relieved by rest, heat, and immobilization in a plaster cast. Chronic disability requires arthrodesis of the talonavicular, the talocalcaneal and the calcaneocuboid joints. The valgus of the heel must be corrected and the navicular properly aligned with the head of the talus. Operation is postponed until adequate bony development of the tarsus has been attained.

Genuine Peroneal Spastic Flatfoot. Acute spasm of the peronei occurs reflexly from an irritative focus within the tarsal area. Most often it is due to rheumatoid arthritis, although it can be associated with an acute sprain, fracture, degenerative arthritis, infection, etc. Pain occurs over the peroneal area where the tendons stand out prominently beneath the skin. The foot is pulled into valgus, and any attempt to stand or walk greatly intensifies the pain and causes flattening of the longitudinal arch. Spasm can be demonstrated by electromyographic studies or under anesthesia when the valgus deformity disappears spontaneously. Heat, immobilization and sedation control the painful spasm. The attack is directed to the causative lesion.

OSTEOCHONDROSIS OF THE TARSAL NAVICULAR
(Köhler's Disease)

This condition is characterized by aseptic necrosis of the navicular bone, causing a painful limp in a child.

The navicular (scaphoid) is always the last



Fig. 461. Köhler's disease or osteochondrosis of the tarsal scaphoid.

tarsal bone to ossify. The average age of appearance of the center of ossification is between 18 months and 2 years in girls and between 2 years and 3 years in boys. Normally, development of ossification is quite variable, and not infrequently the ossific nucleus may be somewhat dense and flattened or fragmented. Such changes may be difficult to distinguish from those of Köhler's disease.

Etiology and Pathogenesis. Pathologic specimens have been rare but have shown scattered areas of aseptic necrosis and absorption of bone.[23] The cause is unknown, but the microscopic picture strongly suggests vascular obstruction as the offender.[24] In the first few years, the cartilaginous navicular is surrounded by a ring network of vessels which

[23] Recine, A.: Le osteochondriti. Relazione al XXXIX Congresso della Società Italiana d Ortopedia e Traumatologia, Napoli, 13-15 ottobre, 1954.

[24] Waugh, W.: The ossification and vascularization of the tarsal navicular and their relation to Köhler's disease, J. Bone & Joint Surg. 40B·765, 1958.

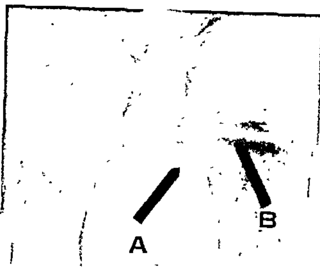


FIG 459. Talocalcaneal bridge. The sustentaculum tali is prominent, and the talocalcaneal articulation is obliterated in its medial portion (Shands, A. R., Jr., and Wentz, I. J.: S. Clin. North America 33:1643)



FIG. 460. Calcaneonavicular bar. Note the spurring at the superior talonavicular articulation and the os subcalcis beneath the calcaneus. (Shands, A. R., Jr., and Wentz, I. J.: S. Clin North America 33:1643)

talocalcaneal bridge forces the calcaneus into valgus, which, when severe, causes valgus at the midtarsal joint, dropping of the longitudinal arch, and a flatfoot.

A less common anomaly is the *calcaneonavicular bar* (synostosis or coalescentia calcaneonavicularis) The bridging from the anterior end of the calcaneus to the navicular may be complete or incomplete. The rare accessory tarsal bone, the calcaneus secundarium, probably represents incomplete bridging Although inversion-eversion subtalar motion is restricted, the calcaneonavicular bar may only occasionally be associated with the deformity of valgoplanus. The bar may be fractured.

SYMPTOMS

Even in the presence of marked deformity there may be no symptoms. This is especially true when a bony bridge is present. However, interference with the mechanics of talonavicular motion inevitably leads to degenerative arthritis of this joint with production of symptoms in adult life.

Symptoms are most common when a severe deformity is due to an incomplete type of juncture. Often between the ages of 12 and 15, a severe wrenching injury tears the juncture or fractures the bony projection, especially from the anterior end of the calcaneus An acute attack of foot pain, swelling and tenderness about the tarsal area may be associated with peroneal muscle spasm Symptoms and findings disappear, and chronic persisting pain and disability eventuate. The history of long-standing deformity may date back to childhood.

FINDINGS

A valgoplanus foot deformity is apparent. The heel is in eversion, and the navicular is displaced laterally so that the head of the talus is prominent on the medial border of the foot. Subtalar motion is very limited or nonexistent. During the acute stage, peroneal spasm is evidenced by pain over taut and prominent tendons in the peroneal area The spasm subsides under anesthesia, at which time the restricted subtalar motion will be found to persist.

Roentgenologic Findings. The talocalcaneal bridge is demonstrated by an axial view of the calcaneus. The x-ray tube behind the heel is directed downward and forward at an angle of 45°. By bending the knees and dorsiflexing the foot at the ankle, interference by soft-tissue shadows is eliminated. When a talocalcaneal bridge is present, the medial side of the subastragalar joint is obliterated. A secondary change seen in the lateral view is marginal lipping on the dorsal aspect of the talonavicular joint.

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Treatment

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This condition is characterized by aseptic necrosis of the navicular bone, causing a painful limp in a child.

The navicular (scaphoid) is always the last tarsal bone to ossify. The average age of appearance of the center of ossification is between 18 months and 2 years in girls and between 2 years and 3 years in boys. Normally, development of ossification is quite variable, and not infrequently the ossific nucleus may be somewhat dense and flattened or fragmented. Such changes may be difficult to distinguish from those of Köhler's disease.



Fig. 461. Köhler's disease or osteochondrosis of the tarsal scaphoid.

Etiology and Pathogenesis. Pathologic specimens have been rare but have shown scattered areas of aseptic necrosis and absorption of bone.[23] The cause is unknown, but the microscopic picture strongly suggests vascular obstruction as the offender.[24] In the first few years, the cartilaginous navicular is surrounded by a ring network of vessels which

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[24] Waugh, W.: The ossification and vascularization of the tarsal navicular and their relation to Köhler's disease, J. Bone & Joint Surg 40B:765, 1958.



FIG. 462. Bipartite sesamoid.

originate from the dorsalis pedis and the medial plantar arteries. From this peripheral network a single nutrient artery penetrates to the center of the cartilaginous structure and becomes surrounded at its tip by developing ossification. Soon other penetrating vessels follow, and multiple small areas of ossification join into a single large ossific nucleus. When the formation of this central arterial supply is delayed, the ossification center may become dependent upon as little as a single artery as late as the 5th year of age. The tremendous compression forces to which the navicular is constantly exposed may compromise the limited vascular supply, and the ossification center undergoes aseptic necrosis. Reactive hyperemia developing about the bone explains the pain, the swelling, the tenderness and the local heat. Ingrowth of new vessels effects resorption and replacement of necrotic bone.

Clinical Picture. The age incidence is 4 to 10, with an average of 5. Both sexes are affected, but the condition is far more common in boys. The child limps and complains of pain in the foot. Slight swelling and tenderness are localized to the navicular, and occasionally a sensation of local heat is perceived

Roentgenographic Findings. The scaphoid displays increased density, loss of trabecular structure and alteration in size and shape Two types of abnormality are seen: (1) the navicular is flattened and displays patchy areas of increased density. The lateral view is comparable with the edge of a disk. Over a period of about 2 years the size and the shape are gradually restored, and the trabecular pattern reappears. (2) The navicular possesses a normal shape but appears to be denser than surrounding bones. Within a few months the dense bone is gradually absorbed until, after about a year, a faint ossific shadow remains. At about 2 years after the onset several small centers of ossification make their appearance, and the bone thereafter is gradually restored. The navicular is always reconstituted to normal, the process taking from 18 months to 3 years.

Treatment. Immobilization of the foot during the acute stage relieves the discomfort but does not appear to hasten the process. A cast may be worn for several weeks and followed with a rigid shank shoe and a Thomas heel. As soon as symptoms are relieved, full activity may be allowed. Complete reossification within 2 to 3 years may be expected.

SESAMOID BONES OF THE FOOT

Where a tendon courses over a bony prominence where it is subjected to friction and pressure, it develops a protective cartilaginous nodule. Some of these nodules ossify during childhood and become sesamoid bones. The function of a sesamoid is to provide an articulating surface and a fulcrum which increases the strength of muscle pull. As an articulating bone, it is subject to degenerative changes and fracture.

The accompanying diagram defines the usual locations for sesamoid bones The two under the first metatarsophalangeal joint are the only ones which are constant. They should not be mistaken for loose joint mice or fracture fragments.

Other common sesamoids are those found in the peroneus longus where it courses around the cuboid, in the tendon of the

tibialis posterior beneath the scaphoid, in the short flexors of the toes beneath the metatarsophalangeal joints, and in the long flexors beneath the interphalangeal joints.

Sesamoid Bones of the Large Toe

A sesamoid bone is invariably situated in each tendon of the flexor hallucis beneath the first metatarsal head. Ossification centers appear between the ages of 8 and 11. When a sesamoid develops from several centers, the bone consists of two or more individual segments, constituting the bipartite, the tripartite, or the multipartite sesamoid. The sesamoids are covered with hyaline cartilage on their dorsal aspect by which they articulate with the cartilage covered under surface of the metatarsal head. They function to (1) provide a fulcrum by which the efficiency of flexion of the large toe is enhanced, and (2) to relieve the metatarsal head from weight-bearing pressure.

The tendon of the flexor hallucis brevis surrounding the sesamoid may be painfully contused by direct trauma such as a fall from a height. Weight-bearing is painful, and tenderness is localized to the site of the sesamoid. Passive dorsiflexion and active plantar flexion of the toe accentuate the discomfort. Disability is temporary, and cure is expedited by rest, abstinence from weight-bearing, heat, needling the tendon under local anesthesia, and placing a felt pad in the shoe behind the *sesamoids.*

The sesamoids, especially the medial, may *be fractured. Pain,* tenderness and swelling are situated over the affected sesamoid. The discomfort is intensified by any movement of the large toe. X-ray films reveal several fragments with jagged osteoporotic margins. In contrast, a *multipartite* sesamoid displays individual portions which are smooth, rounded and completely encircled with cortex; the junctions generally lie transversely. If one is in doubt, the diagnosis is deferred until a sufficient period of immobilization demonstrates the presence or the absence of union between fragments.

Osteoarthritis of the *joint between* the sesamoids and the undersurface of the metatarsal head is manifest by pain on *weight-*bearing, tenderness over the sesamoids, a grating sensation on *movement* of the toe, and stiffness after rest. Recurring incidents of pain is an indication for removal of one or both sesamoids. Removal of both bones results in a painful joint which is poorly adapted to weight-bearing. The metatarsal head must be elevated constantly by a felt pad placed just behind it.

CALLUSES

A callus is a growth composed of hypertrophied horny layer of squamous epithelium in response to abnormally excessive pressure. It is a physiologic defense process. Desquamation and restoration of normal skin follow elimination of pressure. The usual sites for development of a callus are at the points of bony prominences: the plantar aspect of the 1st metatarsal head where sesamoids are situated, the plantar surface over the 2nd metatarsal head, the dorsal aspect and the tips of clawed toes, the dorsum of the foot at the base of the 1st metatarsal, the skin overlying a bunion, and over the os calcis at the insertion of the Achilles tendon. Calluses over the plantar surface of the foot are prone to develop when the transverse arch is flattened by ligamentous inadequacy, a foot deformity which throws the forefoot downward, paralysis of the foot dorsiflexors and intrinsics, and thinning of the subcutaneous fat pad which occurs in old age. Elsewhere the hornified epithelium develops where the skin is compressed between the bony prominence and the shoe. The callus in itself is not painful. As a thickening over a bony prominence, it increases the pressure, and an adventitious bursa forms beneath it. A traumatic bursitis is the cause of the discomfort. Where a callus develops between the toes, perspiration softens the callus, and a soft corn results. An underlying exostosis is the usual cause.

Treatment

Removal of pressure is the essence of treatment. Shoes of adequate width and low heels *are worn. A felt pad is placed* behind or surrounding the bony prominence. The callus is trimmed down flat but should not be completely excised, as a sloughing indolent ulcer *may result.* Hot soapy foot soaks are *prescribed* and followed by application of a keratolytic agent such as a 20 per cent solution of salicylic acid in collodion. Irradiation of the

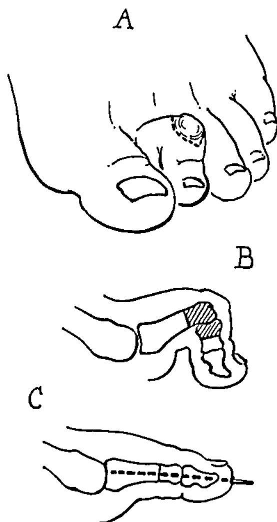


Fig. 463. Hammer toe, surgical correction. (A) Elliptical excision of dorsal callus. This optional step should be eliminated if primary closure will be difficult. The callus will disappear as pressure is eliminated. (B) Extent of bone excision. (C) Corrected position and intramedullary pin fixation.

callus has no effect. Surgical treatment demands correction of foot deformity and removal of the bony prominence. A painful callus over the metatarsal head is cured permanently by resecting the entire head or the condyles on its plantar aspect.

Technic.[25] Through a longitudinal incision over the dorsal aspect of the metatarsophalangeal joint, the extensor tendon is retracted to one side, and the capsule is cut longitudinally. The head of the metatarsal is dislocated dorsally out of the wound while the toe is flexed plantarward. This exposes the sharply pointed condyles which extend from before backward and plantarward. A sharp osteotome is placed at the junction of the condyles with the joint surface and lined up parallel with the metatarsal shaft. Then the condyles are excised. The dislocation is reduced, and the capsule and the skin are sutured. Postoperatively, an anterior metatarsal bar is placed on the shoe. Painless walking is resumed in 3 weeks, and the lesion gradually disappears.

[25] DuVries, H L · New approach to the treatment of intractable verruca plantaris (plantar wart), J.A M A. 152·1202, 1953.



Fig. 464. Hammer toe. Arthrodesis by intramedullary pinning.

WARTS

Plantar warts occur on the ball of the foot but are situated independent of points of pressure. Usually a large "mother wart" is surrounded by several smaller warts The verruca is often imbedded in callus, the center of the lesion being papillary, soft and very vascular. They may disappear and reappear spontaneously. Roentgenotherapy dries up the root of the wart, and the latter drops out. Electrocoagulation and surgical excision should be avoided.

HAMMER TOE

A hammer toe is one which is flexed at the proximal interphalangeal joint so that the tip of the toe points downward. In contrast with a clawed toe, the metatarsophalangeal joint is not hyperextended. The cause seems to be

congenital, because the condition is often bilateral, associated with other defects, and other members of the same family are similarly afflicted. A tightly fitting shoe or a valgus displacement of the large toe frequently initiates and encourages the deformity. A painful bursa and callus overlies the dorsal prominence of the flexed interphalangeal joint. The tip of the toe is broadened and thickened with callus as a result of excessive pressure. The metatarsal head is depressed, and a callus covers its plantar aspect. The second toe is affected most often.

Treatment

Early deformity in childhood may be overcome by adhesive strapping. A strip of tape is passed over the dorsal prominence and fastened beneath the adjacent toes. The mother frequently manipulates the toe into extension. The toe box of the shoe must be adequate.

Severe deformity requires surgical correction. The dorsal callus is removed by an elliptical incision. The head of the proximal phalanx and the base of the disal phalanx are excised, the toe straightened, and an intramedullary pin provides fixation until fusion is complete.

INGROWING TOENAIL

The lateral margins of a toenail or the anterior corners may dig into the surrounding skin and cause a paronychial infection. The main causes are a tightly compressing shoe or trimming the nail too short so that the corners in growing outward penetrate the skin. An infectious granuloma may form.

Conservative Treatment

Hot soapy soaks localize the infection and promote drainage. The corners of the nail should be freed and kept elevated by pledgets of cotton soaked in an antiseptic. Shoes must be of adequate width and low heeled.

Surgical Treatment

The toe is anesthetized by block infiltration about its base. A small incision is made over the eponychium, and the base of the nail over the affected side is exposed. The lateral third of the nail is removed, and the underlying

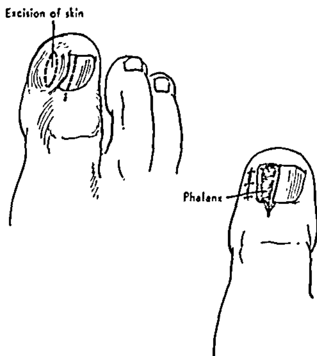


Fig. 465. Ingrown toenail. An elliptical segment of skin is removed, and suturing the defect retracts the skin laterally. The nail must be removed down to the bony surface.

matrix is curetted down to the phalanx. On the lateral side of the toe, an elliptical segment of skin is removed, and the edges are sutured together. This draws the skin away from the edge of the remaining nail. Petrolatum gauze is packed into the wound over the phalanx, and a compression dressing is applied. The toe is bathed in hot soapy soaks for several days without removing the dressing. After 3 days, the pack is removed and is followed by dry dressings.

HALLUX RIGIDUS

Hallux rigidus is a condition of the big toe manifested by painful restricted motion, particularly dorsiflexion, of the metatarsophalangeal joint, and eventual degenerative arthritis.

Clinical Picture

The condition starts as early as late childhood and adolescence, but symptoms may develop at any time from adolescence to old age.

Sex. In adolescence, females are predominantly affected, perhaps because of wearing high-heeled shoes. In adults, the two sexes are equally affected.

Symptoms and Findings in the Adolescent

Pain in the big toe may start insidiously but often is precipitated by injury, such as stubbing the toe. A previously apparently normal toe becomes painful on walking, especially at the point in the step when the toe is dorsiflexed prior to the "push-off." The pain is throbbing or aching and is relieved only by rest. To avoid discomfort, the gait is hopping and short-stepped.

On examination, the metatarsophalangeal joint displays soft-tissue swelling, tenderness, and a position of slight flexion. Tenderness is often most acute over the dorsal aspect. The toe is held immobile by muscle spasm. Passive attempts to dorsiflex the toe result in failure and they cause severe pain, whereas plantar flexion is almost normal. Some crepitus on motion may be noted. The interphalangeal joint is hypermobile, as much as 60° to 70° of passive dorsiflexion being obtainable. The foot is usually long, narrow, flat and pronated. The great toe is often excessively long.

This acute stage subsides and is followed by a prolonged disability of repeated exacerbations and increasing stiffness of the big toe.

Symptoms and Findings in the Adult

Progressively increasing stiffness of the big toe may develop without antecedent pain or subsequent to a prolonged course of repeated attacks of pain and swelling. The symptoms in the joint are typically osteoarthritic and subject to acute exacerbations as a result of an injury such as stubbing the toe or an unguarded movement in taking a step. In the intervals between painful episodes, the joint may be slightly painful or not at all. The protective gait is characteristic.

The hallux rigidus gait is described as follows: In the normal heel-to-toe motion, the weight of the body rolls along the outer side of the foot until the heel is off the ground and the heads of the 4th and the 5th metatarsals are bearing weight; it then passes across the metatarsal head area to the head of the first metatarsal and extends progressively toward the terminal phalanx of the big toe. In hallux rigidus, the first metatarsal head is displaced dorsally, and the metatarsophalangeal joint is in the plantar-flexed position. The body weight in the last phase of the step is transferred laterally to the 2nd and the 3rd metatarsal heads and passes forward to the terminal phalanx of the big toe, bypassing the 1st metatarsophalangeal joint. In consequence, the push-off is accomplished by the outer 4 toes and the distal phalanx of the big toe, thereby protecting the 1st metatarsophalangeal joint from painful dorsiflexion. The flexion of this joint is maintained by spasm of the flexor hallucis brevis. Painful calluses develop beneath the heads of the central 3 metatarsal heads and along the outer border of the heel. A characteristic excessive wear on the shoe occurs under the terminal phalanx of the great toe. The upper of the shoe bulges outward.

The 1st metatarsophalangeal joint in late stages is completely rigid and displays a permanent flexion deformity. A large exostosis on the dorsum of the 1st metatarsal is covered by a painful, tender soft tissue swelling of bursitis. The interphalangeal joint is hypermobile; occasionally, its motion is diminished

Roentgenologic Findings. These are typical of degenerative arthritis. In the adolescent, the roentgenogram is negative.

Pathology[26]

In the earliest stage, erosions occur in the articular cartilage at the center and dorsal margin of the base of the proximal phalanx and the corresponding surface of the metatarsal head. The synovial membrane shows changes consistent with traumatic synovitis The roentgenologic characteristic of increased density and fragmentation of the epiphysis of the proximal phalanx, as described by some authors, is merely coarsely trabeculated compact living bone arising from 2 centers of ossification. It is seen in normal asymptomatic joints.

Late stages show well-marked osteoarthritis throughout the joint. The exostosis develops on the dorsum of the metatarsal head as a response to pressure against the shoe.

Etiology

The actual cause is unknown. Theories which have been advanced are:

[26] Bingold, A. C, and Collins, D. H.: Hallux rigidus, J. Bone & Joint Surg 32B:214, 1950.

1. Hypermobility of the First Metatarsal.[27,28] To provide stability for flexion of the big toe in standing and walking, the 1st metatarsal must be held in a downward pointed direction. When the latter becomes horizontal or dorsally displaced by weak musculature or a poorly fitted shoe, the big toe must exert an abnormal degree of flexion. The dorsal aspect of the metatarsal head becomes nonarticulating and undergoes degenerative changes. In addition, the strong flexion of the toe exerts excessive pressure on the articulating surfaces and causes erosions. The plantar portion of the capsule becomes shortened. Dorsiflexion is restricted. Any attempt to dorsiflex the toe increases the articular compression. A vicious cycle is set up, and a progressive degeneration is the result. The interphalangeal joint undergoes compensatory increased mobility.

2. Trauma and Inflammation. These factors supposedly may initiate a similar vicious cycle, but this theory is questionable in view of the failure to develop a hallux rigidus following such conditions as gout, fracture, etc.

Treatment

Management of the early acute case without severe degenerative arthritis differs from the severe adult case.

Treatment During Adolescence. The acutely painful 1st metatarsophalangeal joint requires rest, heat, sedation to overcome muscle spasm, and the application of continuous traction to the toe. Injection of hydrocortone or meticortelone effects rapid subsidence of pain and swelling. When acute symptoms have subsided, frequent repeated manipulations are necessary to restore full dorsiflexion. Forceful traumatic manipulations, even under anesthesia, are to be deplored. A well-fitted shoe with adequate room in the toe box is worn, and the patient is instructed on proper heel-to-toe walking.

Persistence of symptoms or recurrences are an indication for osteotomizing and depressing the 1st metatarsal, as advocated by Lambrinudi. Its success depends on its performance before degenerative changes have ensued.

Treatment of Advanced Case. The injection of hydrocortone or meticortelone into the joint may control pain satisfactorily, but the abnormal gait, metatarsal pain, and recurrences of pain in the big toe constitute a severe disability. Surgical correction is necessary. Resection of the base of the proximal phalanx (Keller arthroplasty) provides good mobility and freedom from pain, but the power of push-off is weakened. Arthrodesis permanently relieves pain but is least appreciated by women. Osteotomy and depression of the first metatarsal segment may be added, particularly with an arthroplasty.

Metatarsal pain, often associated with hallux rigidus, requires a metatarsal pad or bar, resection of the offending metatarsal heads, or removal of their condyles.

PAIN ABOUT THE HEEL

Pain about the heel may occur from conditions on the posterior and plantar aspects, within the calcaneus itself, and from the talocalcaneal joint. These conditions may be classified as traumatic, infectious, static and neoplastic.

Plantar Calcaneal Spurs and Fascitis

On the plantar aspect of the calcaneus at the attachment of the plantar aponeurosis, a bony prominence may develop and points distally. Actually, the spur extends transversely across the entire plantar surface of the bone. Frequently, the spur is asymptomatic. It is generally thought to develop because of traction on the periosteum with consequent subperiosteal ossification. The cause of pain is not well understood. Theories advanced include: (1) inflammation of an overlying bursa; (2) traumatic periostitis; (3) tearing or strain of the fibers of attachment of the fascia; (4) focal sepsis causing localized inflammation.

Clinical Picture. Pain on the undersurface of the heel occurs on standing and walking and is relieved by rest. Localized tenderness is found, particularly over the medial portion of the spur. Slight swelling may be observed. Passive dorsiflexion of the toes may accentuate the discomfort.

[27] Jack, E. A.: The etiology of hallux rigidus, Brit. J. Surg. 27:492, 1940.

[28] Lambrinudi, C.: Metatarsus primus elevatus, Proc. Roy. Soc. Med. 31:1273, 1938.

Roentgenologic Findings. A spur may or may not be visible.

Treatment. *Conservative* treatment includes bed rest, hot soaks, needling the tender area with a local anesthetic, injection of hydrocortone, removal of foci of infection, and wearing a sponge-rubber heel pad, with the concavity cut out to correspond to the tender area.

Surgical treament for resistant cases includes removal of the spur, stripping the soft tissues from the plantar surface of the calcaneus, and resection of a portion of the plantar aponeurosis at its origin.

Painful Heel Pad[29]

An elastic adipose tissue covers the plantar aspect of the calcaneus. Similar tissue is found at other regions subjected to pressure, e.g., over the ischial tuberosity, the fingertips and the infrapatellar area. The calcaneal fat pad is composed of elastic fibrous tissue and closely packed fat cells These are enclosed in rigid compartments formed by septa of fibrous tissue which extend from the undersurface of the calcaneus to the subcutaneous tissue. Each septum is shaped like a test tube, the "open end" attached to the bone, the "closed end" is observed as a curved loop in the subcutaneous tissue. The effect of this structure is to permit the walls to bulge outward under the influence of pressure, then to spring back promptly before the impact of the next step is received. All areas of elastic adipose tissue are present at birth. It does not develop in postnatal life.

With advancing age, all collagenous tissue degenerates. The heel-pad septa are no exception. Gradually, under the influence of ordinary weight-bearing or suddenly due to a severe impact, the fibrous strands rupture, and the fat cells are spilled Increasing pressures are brought to bear upon the calcaneus, which reacts by increasing density of the cortex and bony proliferation at the margins of the tuberosity. The condition occurs after 40, usually in the obese, and after severe trauma as a fractured calcaneus

Clinical Picture. Pain is experienced beneath the heel, chiefly on standing, less so on walking, and is relieved by rest. The posterior weight-bearing portion of the tuberosity is generally tender, in contrast with the localized tenderness of a spur found more anteriorly.

Roentgenologic Findings. *The normal calcaneal fat pad*, as revealed by soft-tissue exposures, displays (1) a dense layer of fibrous tissue covering (2) the smooth undersurface of the calcaneus, (3) the U-systems, which are dense, regularly arranged, vertically disposed strands with subcutaneus loops.

The *abnormal calcaneal fat pad* reveals (1) bony proliferation about the margin of the tuberosity, (2) decreased density of the underlying fibrous layer, (3) elastic adipose tissue is less dense, (4) strands are thinner, irregular, broken in outline, not parallel, and (5) the U-loops are distorted.

Treatment is palliative. Removal of pressure relieves the acute discomfort. Weight-bearing pressure is reduced by a soft rubber heel pad with concavity scooped out, wearing sponge-rubber heeled shoes, and in women a high heel with an inclined heel platform will transfer the weight anteriorly.

Subastragalar Arthritis

Most commonly, this is a degenerative arthritis which is secondary to trauma. The failure to observe a fracture on the x-ray films does not rule out the possibility of tears in the articular cartilage with small crack fractures through the articular cortex. In time, increasing sclerosis, irregularity, and narrowing of the joint become apparent. An irritative synovitis often is the first manifestation of a lesion, infectious or neoplastic, of the talus or the calcaneus.

Clinical Picture. Pain and swelling about the hind part of the foot are usually most pronounced, with weight-bearing; relief is obtained by rest. The heel is often held in eversion by reflex peroneal spasm, attempts at passive inversion are painful, and the longitudinal arch is flattened. Deep pressure over the sinus tarsi in front of the lateral malleolus evokes tenderness. Occasionally, crepitus is palpable on inversion-eversion movement. Injection of a local anesthetic into the sinus tarsi localizes the point of origin of pain.

Roentgenologic Findings. Oblique and postero-anterior projections reveal typical findings of degenerative arthritis in the talocalcaneal joint. Osteoporosis and loss of joint outline should suggest infection or neoplasm

[29] Kuhns, J. G · Changes in elastic adipose tissue, J. Bone & Joint Surg 31A 541, 1949.

Treatment. Subastragalar arthritis of traumatic origin constitutes a chronic disability which often resists all forms of conservative treatment. A triple arthrodesis should be done.

Calcaneal Epiphysitis (Sever's Disease)

The posterior portion of the calcaneus develops as an independent center of ossification, separated from the main bone by a cartilaginous epiphyseal plate. The center appears at age 10 and fuses with the rest of the bone at about 15. The epiphyseal plate is vertically disposed and therefore subjected, by calf muscle pull, to strong shearing stresses. During the rapid growth period of puberty, the epiphyseal-diaphyseal juncture is weakened by an excess of fragile calcified cartilage. Multiple microscopic fracture separations occur, circulation to the epiphysis is cut off temporarily, the epiphyseal bone becomes necrotic and appears to be relatively dense. Eventually, granulation tissue loops from the parent bone penetrate the epiphysis, resorbing the dead bone, which now appears as multiple fragments, and reconstructing by creeping substitution plus endochondral ossification. This theory satisfactorily explains the clinical picture.

Clinical Picture. The condition is most common between the ages of 11 and 14. Males, probably because they are more athletic, are predisposed. A history is often given of running hard or a long hike. The onset of pain and limp is gradual. Pain is situated over the epiphysis, is aggravated by wearing low heels, standing on tiptoe, and pressure of a rigid shoe counter. It is relieved by relaxing the calf muscles by resting with the knee flexed and the foot in equinus. The patient walks with short halting steps to avoid dorsiflexion at the ankle. Tenderness is well localized over the lateral and the medial sides, anterior to the tendocalcaneus attachment, where the epiphyseal plate comes to the surface.

Roentgenologic Findings. The epiphysis appears to be denser than the main bone, which is somewhat rarefied by reactive engorgement. At a later stage, fragmentation of the epiphysis indicates active resorption and replacement. The epiphyseal line is cloudy and ill-defined. Premature epiphyseal closure may occur.

Many authors regard increased density and fragmentation as normal, because these are frequently found in individuals without symptoms. However, symptoms are due to epiphyseal-diaphyseal separation, last but a short time and are often overlooked; whereas the changes of reconstruction proceed asymptomatically over a long period of time.

Treatment. Although symptoms are short-lived, recurrences are probable until the epiphyseal line is obliterated. The aim of treatment is the relief of tension from the epiphysis. If pain is severe, a thigh-length cast is applied with the knee in flexion and the foot in equinus. If discomfort is less severe, all athletic activities are curtailed, the foot is held in equinus by an elastic bandage, adhesive strapping, or a heel elevation, and, if unilateral, crutches are used. Physical therapy is of no value.

Tenosynovitis of Achilles Tendon

Tenosynovitis most often is a result of excessive use of the calf muscles. The sheath is swollen, distended by an effusion, tender, and a fine crepitus is perceived with motion. Treatment consists of immobilization and avoiding the provocative occupational or athletic activity. Rarely, suppurative tenosynovitis may be initiated by a wound. Symptoms included severe pain, marked swelling, heat and acute tenderness over the tendon. The foot is held in equinus to relax the tendon. Passive dorsiflexion intensifies the pain. Constitutional symptoms include fever, chills, leukocytosis, etc. Treatment demands immediate open drainage and antibiotics.

Achilles Bursitis

Above the point of insertion of the tendon, a small interval between tendon and bone is occupied by a bursa. Bursitis is manifest by pain at this site intensified by dorsiflexion of the foot. Tenderness is situated behind the tendon.

At the posterosuperior angle of the calcaneus, a bony prominence often develops in response to repeated pressure by the counter of the shoe. An adventitious bursa develops superficial to the tendon in the subcutaneous area. Pain, tenderness and swelling are easily detected as being just beneath the skin. Simi-

Roentgenologic Findings. A spur may or may not be visible.

Treatment. *Conservative* treatment includes bed rest, hot soaks, needling the tender area with a local anesthetic, injection of hydrocortone, removal of foci of infection, and wearing a sponge-rubber heel pad, with the concavity cut out to correspond to the tender area.

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[29] Kuhns, J. G. Changes in elastic adipose tissue, J. Bone & Joint Surg 31A.541, 1949.

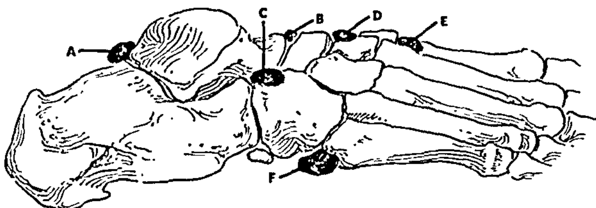


Fig. 467. Accessory bones of foot seen from fibular aspect. (A) Os trigonum. (B) Dorsal talonavicular ossicle. (C) Os calcaneus secundarius. (D) Os intercuneiforme. (E) Os intermetatarseum. (F) Os vesalii. (After Wood Jones)

tendons. The actual cause is unknown, but histologic findings point to a chronic inflammatory process.

Gross Pathology

The plantar fascia is thickened by formation of a lobulated, firm, irregular mass of smaller fused nodules. The cut section shows gray fibrous tissue arranged in fine bands and whorls, or sharply circumscribed zones of dull grayish-yellow tissue. The growth is often located in the longitudinal arch and less often beneath the metatarsal heads. The process does not tend to involve adjacent structures, but the growth is often adherent to subjacent muscles, tendons, nerves and blood vessels and to the overlying skin.

Histopathology

The nodules consist of highly cellular islands of proliferating fibroblasts which contrast sharply with an acellular collagenous background. The nodules are discrete or merge gradually with surrounding fascial bundles The typical fibroblasts contain elongated or oval nuclei with scant vesiculated chromatin and one or two small pink nucleoli Loose collagenous fibers are distributed between the fibroblasts. The fibroblasts are uniform, but rarely they display slight variations in size, shape and staining reactions and occasional mitotic figures Some nodules are relatively acellular and consist of dense collagen A constant finding within the proliferative nodules is the formation of blood vessels lined with hobnail-like endothelial cells and surrounded by plump fibroblasts. Inflammatory cells consisting chiefly of lymphocytes and mononuclears are situated at a distance from the proliferative nodules.

Clinical Picture

A hard tender fixed mass develops in the sole, usually in the longitudinal arch. It is adherent to the overlying skin, which appears wrinkled and indented about the swelling. Inflammatory signs are absent. The growth slowly enlarges over a period varying from months to years, and eventually pain on standing and walking is complained of. Other evidence of the fibroblastic tendency may be found, and a history of epilepsy is often obtained.

Treatment

The mass and the plantar aponeurosis must be excised. The sole may be laid open by an incision which runs longitudinally along the medial or the lateral border of the foot and extends transversely beyond the metatarsal heads. Incisions at the middle of the sole, especially if made longitudinally, will almost surely produce a keloid. Recurrences are common, but the lesion is benign.

ACCESSORY BONES OF THE FOOT

Extra, small bones are occasionally found in the foot Each originates from a separate ossification center of a tarsal or a metatarsal bone which fails to fuse with the main bone. Instead, the accessory bone forms a fibrous or cartilaginous junction with the parent bone. Rarely, a synovial-lined cavity intervenes. When the secondary center of ossification becomes fused by osseous tissue to the main

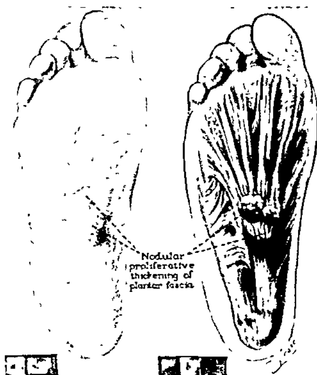


Fig. 466. Plantar fibromatosis. Nodular fibrous thickenings develop in the plantar aponeurosis, particularly in the longitudinal arch. (Meyerding, H., and Shellito, J. G.: Dupuytren's contracture of the foot, J. Internat. Coll. Surgeons 11:595)

lar bursae may develop over any bony protrusion subjected to pressure and friction.

Treatment. The aim of treatment is elimination of pressure. The counter is cut away, or an open-back shoe is worn. Excision of the bony excrescence may be indicated, especially for men. Acute symptoms are alleviated by puncturing the bursa, injecting hydrocortisone, and hot soaks. Excision of the bursa is never indicated. Unless the causative factor is removed it will recur.

Exostosis Below Lateral Malleolus

Compression fracture of the os calcis often causes lateral spreading of the bone. The bony prominence impinges against the lateral malleolus during eversion movement. Standing and walking are painful. Tenderness is perceived immediately below the malleolus. The condition is often confused with subtalar arthritis, but an axial view of the calcaneus reveals the bony mass, and injection of a local anesthetic beneath the malleolar tip relieves the discomfort and defines the cause. Excision of the excess bone is indicated.

Xanthoma of the Achilles Tendon

A very indurated, indolent, nontender, slowly growing mass may develop in the tendon, particularly at its insertion. Clinically, the tendo achilles has the appearance of being markedly hypertrophied, the mass ending abruptly at about the middle of the posterior aspect of the heel. The tumor has a ligneous, lobulated feel but is not painful and apparently does not interfere with ankle motion. Grossly, the tumor lies on and is imbedded in the tendon. Although localized, it is freed with difficulty. The cut surface is gray but flecked with streaks of yellow pigment.

Microscopically, the tumor consists of groups of polygonal cells, containing cholesterol fat with carotene, and lying in an abundant fibrous stroma. Occasional multinucleated giant cells are seen. The degree of yellow coloring depends on the amount of pigment.

Treatment. The mass is benign and grows very slowly. Excision is not followed by recurrence.

Tumors and Infections

These must definitely be considered in a differential diagnosis of pain in the heel. (For characteristics, see appropriate sections on "Bone Tumors" and "Infections.")

PLANTAR FIBROMATOSIS
(Dupuytren's Contracture of the Foot)[30, 31]

Fibrous nodules may develop in the plantar aponeurosis in a manner similar to Dupuytren's contracture of the palmar aponeurosis The lesion is often interpreted as a fibrosarcoma, but typical histologic characteristics and course identify it as a distinct and benign condition.

Etiology

The growth may develop at any age but frequently at middle age or beyond. Epileptics are predisposed. Often, associated conditions include other manifestations of a fibroblastic tendency such as Dupuytren's contracture, Peyronie's disease, knuckle pads, periarthrosis humeri, keloids, and fibrous nodules in

[30] Meyerding, H, and Shellito, J. G.: Dupuytren's contracture of the foot, J Int. Coll. Surgeons 11.596, 1948.

[31] Allen, R. A., Woolner, L. B., and Ghormley, R. K: Soft tissue tumors of the sole, J. Bone & Joint Surg. 37A:14, 1955.

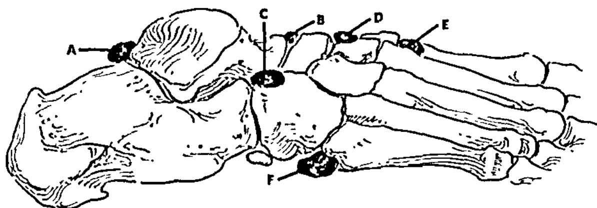


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Treatment

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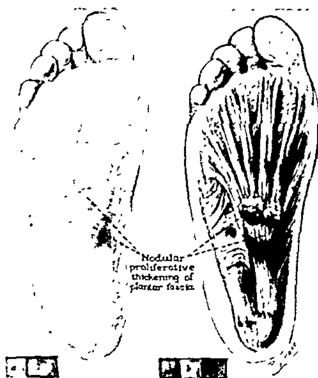


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Fig. 470 (*Top*) Os vesalianum. Its presence bilaterally distinguishes this from an avulsion fracture.

Fig. 471 (*Bottom*) Os supranavicular. The large fragment of bone lying superior to the normal-appearing navicular should be bilateral in order to qualify as an accessory bone. (Shands, A. R., Jr., and Wentz, I. J.: S Clin. North America 33:1643)



Fig. 472. Osteochondrosis of the left (1st) metatarsal epiphysis. It cannot be regarded as an accessory bone. (Shands, A. R., Jr., and Wentz, I. J.: S. Clin. North America 33:1643)

ing the foot against resistance Painful bursae and calluses are caused by friction and pressure of the shoe against the medial bony prominence. If the excess bone impinges against the medial malleolus, limited inversion results The accessory scaphoid must be removed, the remaining bone rounded off, and the posterior tibial tendon transplanted beneath the bone. (See Flatfoot)

2. **Os Trigonum.** A process may develop posteriorly from the body of the talus as the *trigonal process.* This bony extension may be fractured, during plantar flexion at the ankle, by compression between the calcaneus and the posterior lip of the tibia The small posterior fragment is identical in position with the os trigonum, which appears to be separated from but actually is joined by fibrous tissue or cartilage to the talus. The distinguishing characteristics between a fragment and an accessory bone have been described. In addition, in the case of a fracture, air injected into the subastragalar joint will find its way through the fracture gap into the retro-achilles compartment. An os trigonum may be injured by a similar mechanism. Pain over the posterior aspect of the ankle on plantar flexion of the foot is typical. The fragment or the accessory bone must be excised.

3. **Os Vesalii.** The proximal end of the 5th metatarsal often develops as an epiphysis joined by a cartilaginous plate to the main bone The ossification center appears between the 10th and the 12th years and fuses to the shaft at about age 15. Occasionally, failure of fusion occurs, probably because the line of junction is fibrous or because of the constant pull of the attached tendons (peronei brevis and tertius). The separate ossicle constitutes an accessory bone, the os vesalii. The syndesmosis is subject to repeated minor strains. The condition must not be confused with an avulsed fragment, which is always unilateral and eventually unites to the parent bone.

4. **Calcaneus Secundarius.** Incomplete segmentation of the tarsals in the course of development may result in bony bridging between the anterior, superior and medial end

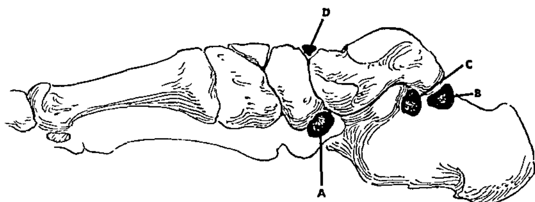


FIG. 468. Accessory bones of the foot seen from the tibial aspect. (A) Os tibiale externum (B) Os trigonum. (C) Os sustentaculi. (D) Dorsal talo-navicular ossicle (After Wood Jones)

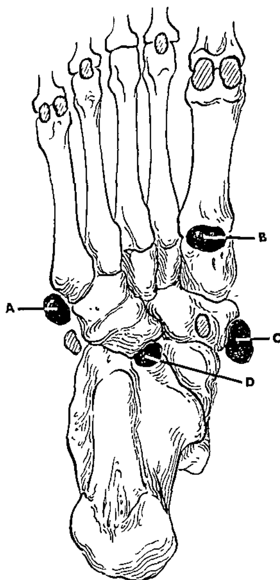


FIG. 469 Accessory bones of the foot seen from the plantar aspect (A) Os Vesalii. (B) Pars peronea metatarsalis. (C) Os tibiale externum. (D) Cuboides secundarius (After Wood Jones)

bone, the latter in the adult state is unusually large and possesses a prominence which represents the accessory center. This is well seen in the scaphoid which often sends medially a large tubercle to which the posterior tibial tendon is inserted. The importance of accessory bones lies in differentiating them from fracture fragments. However, when a bony prominence has developed instead of an accessory bone, it is liable to fracture, and the small fragment lies in the position normally occupied by an accessory bone. An accessory bone possesses smooth borders and is completely encircled by cortical bone. A fracture will eventually show signs of callus formation and obliteration of the fracture line.

An accessory bone may be avulsed from its bone attachment by strong muscle pull. Clinically, this is manifest by severe pain, tenderness, swelling and ecchymosis. A loose fragment may be palpable. Healing takes place by fibrous union.

Accessory bones are often, but not invariably, bilateral. The following are the most important clinically:

1. **Accessory Scaphoid (Prehallux; Os Tibiale Externum; Naviculare Secundarium).** This is the most common accessory bone It appears between the 10th and the 12th years as an ossicle which seems to continue medially from the scaphoid, interrupted only by a vertical line of translucency. The posterior tibial tendon often inserts into the accessory scaphoid, and flatfoot is often associated. Pain and tenderness localized to this site indicate a strain of the syndesmosis or synchondrosis. This is confirmed by pain caused by invert-



Fig. 470 (*Top*) Os vesalianum. Its presence bilaterally distinguishes this from an avulsion fracture.

Fig. 471 (*Bottom*) Os supranavicular. The large fragment of bone lying superior to the normal-appearing navicular should be bilateral in order to qualify as an accessory bone. (Shands, A R., Jr., and Wentz, I. J.: S. Clin. North America 33:1643)



Fig. 472. Osteochondrosis of the left (1st) metatarsal epiphysis. It cannot be regarded as an accessory bone. (Shands, A. R., Jr., and Wentz, I. J.: S. Clin. North America 33:1643)

ing the foot against resistance. Painful bursae and calluses are caused by friction and pressure of the shoe against the medial bony prominence. If the excess bone impinges against the medial malleolus, limited inversion results. The accessory scaphoid must be removed, the remaining bone rounded off, and the posterior tibial tendon transplanted beneath the bone. (See Flatfoot)

2. **Os Trigonum.** A process may develop posteriorly from the body of the talus as the *trigonal process*. This bony extension may be fractured, during plantar flexion at the ankle, by compression between the calcaneus and the posterior lip of the tibia. The small posterior fragment is identical in position with the os trigonum, which appears to be separated from but actually is joined by fibrous tissue or cartilage to the talus. The distinguishing characteristics between a fragment and an accessory bone have been described. In addition, in the case of a fracture, air injected into the subastragalar joint will find its way through the fracture gap into the retro-achilles compartment. An os trigonum may be injured by a similar mechanism. Pain over the posterior aspect of the ankle on plantar flexion of the foot is typical. The fragment or the accessory bone must be excised.

3. **Os Vesalii.** The proximal end of the 5th metatarsal often develops as an epiphysis joined by a cartilaginous plate to the main bone. The ossification center appears between the 10th and the 12th years and fuses to the shaft at about age 15. Occasionally, failure of fusion occurs, probably because the line of junction is fibrous or because of the constant pull of the attached tendons (peronei brevis and tertius). The separate ossicle constitutes an accessory bone, the os vesalii. The syndesmosis is subject to repeated minor strains. The condition must not be confused with an avulsed fragment, which is always unilateral and eventually unites to the parent bone.

4. **Calcaneus Secundarius.** Incomplete segmentation of the tarsals in the course of development may result in bony bridging between the anterior, superior and medial end



Fig. 473. Os calcaneum secundarium. A similar jagged fragment occurring unilaterally must be regarded as a fracture. (Shands, A. R., Jr., and Wentz, I. J.: S. Clin. North America 33:1643)

of the calcaneus, and the inferior, posterior and lateral end of the navicular. (See Calcaneonavicular Bridge.) When cleavage has progressed further, a remnant of this bridge exists as a bony outgrowth from the anterior end of the os calcis, attached by fibrous tissue or cartilage to the navicular. More commonly, a small segment of bone lies within the soft-tissue bridge between calcaneus and navicular and constitutes the calcaneus secundarius. It must be differentiated from a fracture of an anterior calcaneal process. An accessory bone, an anterior process, and complete calcaneonavicular bridge all represent degrees of the same developmental abnormality which causes a rigid, valgus flatfoot.

5. **Os Subfibula and Os Subtibiale.** Small ossicles develop below the outer and the inner malleoli of the ankle. Their fibrous junction with the main bone is insecure, and patients complain of repeated incidents of giving-way and strains. The condition is almost invariably bilateral.

Other less common accessory bones are:

6. **Os Supranavicular**
7. **Os Sustentaculi**
8. **Pars Peronea Metatarsalis Primi**
9. **Os Cuneiform Primum Bipartum**
10. **Os Intermetatarsum**
11. **Os Intercuneiforme et Os Paracuneiforme**
12. **Cuboides Secundarius**

27

The Back

SURGICAL ANATOMY OF THE NERVE ROOTS IN THE LOWER LUMBAR SPINE

The spinal cord terminates in a bulbous enlargement, the conus medullaris at the level of the first lumbar vertebra. It is fixed by a fibrous band which attaches to its terminal end and passes distally through the sacral canal to the coccyx. Nerve roots to the lumbar and the sacral regions pass distally as the cauda equina. The spinal cord is enveloped by the dura, the arachnoid and the pia mater. The epidural space is filled by a thin layer of fat, areolar tissue and veins. Spinal fluid circulates in the arachnoid and the subarachnoid spaces. The pia mater is a thin membrane which intimately surrounds the spinal cord and has lateral extensions, the dentate ligaments, which attach to the inner surface of the dura. These are equally spaced between the nerve roots.

The dura extends as far as the 1st or the 2nd sacral segment The lumbar plexus is formed by the anterior divisions of the 1st, the 2nd, the 3rd and the 4th lumbar nerves. The sacral plexus, often called the lumbosacral plexus, originates from the 4th and the 5th lumbar and the 1st, the 2nd and the 3rd sacral nerves

Each pair of anterior and posterior nerve roots passes to and through the intervertebral foramen enclosed by a dural sleeve. Before they join to form a spinal nerve at the outer margin of the foramen, a bulbous enlargement, the posterior root ganglion, forms and identifies the posterior root. At the lumbosacral level, the first sacral nerve roots are already enclosed in a dural sleeve and are observed as a structure adjacent to and separate from the main dural sac. At the extreme lateral aspect of the spinal canal, the 5th lumbar nerve root is identified where it goes out through the foramen. At the 4th lumbar interspace, the 5th lumbar nerve roots lie within the dural sac on its outermost aspect where it is subject to compression. The only separate nerve structure is the 4th lumbar nerve roots, enclosed in a common dural sleeve, at the extreme lateral aspect of the canal going out through the foramen. The same relationship is observed at the upper lumbar interspaces.

The intervertebral foramen[1] may be divided into two parts. The upper half is bounded above by the pedicle, in front by the body of the vertebra, and behind by the pars interarticularis of the lamina. It is demarcated from the lower half by the slightly prominent lower border of the vertebral body and is occupied by the emerging nerve root. The lower half is bounded in front by the disk, below by the pedicle, and behind by the apophyseal joint. It is occupied only by fibrofatty tissue and small vessels. The nerve enters the foramen by curving around the inferomedial border of the pedicle with which it is in close contact. It then proceeds directly and laterally in a groove on the back of the vertebral body bounded above by the pedicle and the base of the transverse process and below by the rounded margin of the body. At the extreme lateral border of the vertebra, it turns downward and forward to enter the psoas muscle. Except at this extreme lateral edge, the nerve is confined entirely to the upper half of the foramen and is never a direct relation of either the disk or the apophyseal joint. Therefore, a ruptured disk can encroach upon a nerve root only when it lies more medially and goes out at the next space below. For example, a protruded disk at the 4th lumbar space involves the 5th lumbar nerve roots. When an old ruptured

[1] Adkins, E W. O · Spondylolisthesis, J. Bone & Joint Surg. 37B:48, 1955.

disk is scarred and the posterior margins of the contiguous vertebrae develop osteophytic outgrowths, the groove occupied by the nerve root is deepened, but rarely is the nerve root constricted.

REFERRED PAIN FROM THE BACK

Kellgren made studies on the problem of referred pain from the back.[2, 3] He produced consistent patterns of pain and tenderness in various sites by injecting a 6 per cent solution of sodium chloride into various structures. The intensity of pain rose rapidly and subsided in several minutes. Before a structure was injected, the overlying superficial structures were anesthetized with procaine. Then the pattern of pain was produced by merely inserting the needle into the structure, but injection of hypertonic saline was necessary to prolong the pain.

Injection of superficial structures, such as superficial fascia, ligaments, tendon sheaths and superficial periosteum, caused only local pain. Deeper structures, such as muscles, deep fascia, deep ligaments and deep periosteum, when injected, produced a diffuse pain following a spinal segmental pattern, often associated with deep tenderness. Cutaneous sensation was always preserved.

Examples given of pain radiating from the deep structures are pertinent to identifying the origin of a referred pain in an area at a distance When the subscapular area below the scapular spine is injected, pain is experienced about the deltoid region Injection of the base of the first lumbar spinous process causes pain at the iliac crest. Injections deep to the interspinous ligament on one side causes, from L1, pain at the iliac crest, the groin and the scrotum; at L2, pain below the crest and the groin and into the scrotum; at L3, pain in the upper half of the anterior thigh; at L4, pain in the upper lateral ⅔ of the thigh and the posterior lower ⅓ of the thigh and the inner aspect of the ankle; at L5, the same area, but also the entire lateral aspect of the leg; at S1 and S2, pain down the lateral and inner aspect of the thigh to the front of the knee, the posterior thigh, the leg, the heel and the sole of the foot.

The importance of this work lies in the fact that radiating pain must not necessarily be construed as originating from nerve root compression. The deep structures of the back, exclusive of the spine, must be searched diligently for a causative lesion, particularly when no change in cutaneous sensation and muscle weakness exists. The area in which pain is experienced should indicate the approximate location of the offending structure.

THE INTERVERTEBRAL DISK

The intervertebral disks are fibrocartilaginous structures which lie between and separate opposing end surfaces of the bodies of vertebrae. They function chiefly as shock absorbers. An intervertebral disk is affected by progressive degenerative changes, trauma and disease.

Anatomy

Normally, 23 disks exist throughout the spine, being absent only at the atlanto-axial articulation. They are thinnest in the thoracic region and thickest in the lumbar. Each disk is interposed between the bodies of a pair of vertebrae. Each body is composed of cancellous bone covered superiorly and inferiorly by a thin end plate of bone which is perforated by numerous tiny holes. At the periphery of the end plate is a bony thickening, the epiphyseal ring (the Randleiste of Schmorl), which is deficient posteriorly. The end plate is covered by a layer of hyaline cartilage, which may be considered as the outermost portion of the disk. Anteriorly and laterally, the vertebral bodies are bound together by the anterior longitudinal ligament, a strong structure which attaches to their margins and is intimately blended with the underlying disk. Posteriorly, the vertebral bodies are joined by the posterior longitudinal ligament, a weak structure lying within the spinal canal.

Grossly, each disk is composed of a semiliquid central portion, the nucleus pulposus; 2 cartilage plates which separate the nucleus from the bodies of the vertebrae above and below; and a thick ring of fibrous tissue, the annulus fibrosus (lamellosus). The annulus

[2] Kellgren, J H · Observations on referred pain arising from muscle, Clin. Sc 3 175, 1938

[3] ——— · On the distribution of pain arising from deep somatic structures with charts of segmental pain areas, Clin. Sc. 4.35, 1939.

fibrosus arises from the cartilaginous plates, surrounds the nucleus pulposus and inserts into the anterior and the posterior longitudinal ligaments and into the bone of the vertebrae. It is thickest and strongest anteriorly and laterally, being intimately attached to the strong anterior longitudinal ligament. Posteriorly, it is thin and loosely attached to the weak posterior longitudinal ligament. In fetal life small blood vessels penetrate the annulus from the vertebrae, but these soon disappear after birth, and thereafter the disk is avascular. Large bundles of nerves have been demonstrated outside the anterior and the posterior longitudinal ligaments but not within the disk itself.[4]

Microscopically, the cartilaginous plate of hyaline cartilage is situated over the perforated bony end plate but not over the epiphyseal ring. It is cemented to the underlying bone by a thin layer of calcium. It gives origin to the fibrocartilage of the annulus. The annulus fibrosus is composed of alternating layers of fibrous and fibrocartilaginous lamellae. Peripherally, these lamellae interlace and provide firm fixation and resist outward bulging of the nucleus. More centrally the lamellae are horizontally disposed. The fibers of the annulus are attached to (1) the hyaline cartilage, (2) the anterior and the posterior longitudinal ligaments and (3) into the edges of the vertebral bodies, like Sharpey's fibers. The nucleus pulposus is a white glistening body formed of loose, wavy, fibrous strands in a gelatinous and mucoid reticulum. It stains pale blue or pale pink with hematoxylin and eosin. Notochordal cells are said to be present at birth. With advancing age, the water content of the nucleus lessens; fibrous tissue, cartilage cells and amorphous material increase; and the nucleus becomes granular and friable.

Embryology

Vertebrae, cartilaginous plate and annulus fibrosus are derived from mesoderm, nucleus from endoderm being a part of the notochord Provertebrae form and are marked off by the intersegmental artery. Each provertebra divides horizontally, and the caudad half unites with the cephalad half of the adjacent provertebra. Water leaves the cells except in the region of the disk, probably because this region lies furthest from the intersegmental artery.

The notochord becomes extruded into the intervertebral regions, and its canal gradually closes. At the end of the 10th embryonic week, the cells of the vertebral bodies become cartilaginous, and the cells of the notochord lie entirely within the disk. These cells undergo mucoid degeneration. The primitive notochord is the anlage of the nucleus pulposus.

Ossification of the vertebral bodies takes place after the 10th fetal week. Ossification centers for the bodies and each half of the arch appear. Clefts or grooves form in radial fashion on the superior and the inferior surfaces of the bodies. These gradually increase in size for about 10 years, then gradually smooth out by the 25th year. The cartilaginous plate is strongly attached to the disk and extends to the margins of the vertebral body where it is intimately fused with the bony clefts and ridges. On the surface of the cartilaginous plate next to the bone, the process of endochondral longitudinal growth is seen.

The peripheral bone ring or epiphysis is at first a small triangular ring of cartilage which surrounds the superior and the inferior brims of the vertebral body. Small foci of calcium develop in the ring; they ossify, enlarge and fuse to form a bony ring. It is complete at 12 years of age, starts fusing with the body at 15 and completes fusion at the age of 25. Fusion is slowest in the lumbar region. (See the description in section on "Embryology.")

Physiology

The disk serves as an articulation (amphiarthrosis) which gives the spine its mobility. It acts as a cushion or shock absorber. Following loss of a disk, the vertebral body reacts to abnormal pressure forces by hypertrophic bone formation at the surface, revealed as sclerosis and osteophyte formation.

The cartilaginous plate acts as a barrier between the active pressure of the nucleus pulposus and the vertebral body. Like cartilage elsewhere, it degenerates with advancing age, and the nucleus may bulge the bony end

[4] Hirsch, C., and Schajowicz, F Studies on structural changes in the lumbar annulus fibrosus, Acta Orthop scandinav 22:184, 1953

plate inward, causing a characteristic biconcave vertebral body. The nucleus is under constant tension, as evidenced by its bulging outward when the disk is sectioned transversely. If the bony end plate is broken, some of the semiliquid nucleus material is forced into the cancellous bone of the body where it is either absorbed or is encircled by reactive bone and becomes a *Schmorl's node*.

There is *considerable internal pressure* or turgor within the disk. This tends to spread

to ¾ inch taller upon arising in the morning than after *being up all day*. It is probable that *the intermittent* pressure stimulates circulation of tissue fluids through the articular cartilage and thereby nourishes the disk.

PATHOLOGY

The intervertebral disk is subject to continuous and progressive degenerative changes throughout life.[5] These wear-and-tear processes are brought on earlier by excessive functional strain. The earliest changes occur in the nucleus, which in the third decade becomes swollen and granular. The nucleus loses its firmness and glistening appearance and becomes friable and dull gray in color. Later there is destruction of cells and fibers, and the nucleus is converted into a sodden mass. The gel structure is lost. The process extends outward to the annulus, which displays concentric cracks and cavities as early as the 15th year.[6] The annulus becomes progressively weaker and thinner and may be reduced to a thin fibrous ring which may be torn with relatively little force. More commonly, the posterior fibers are stretched by the turgor of the nucleus, and the disk bulges backward into the floor of the spinal canal. A brown pigment of unknown origin is deposited in the annulus, the process of degeneration being known as *brown degeneration*. This fissuring and gradual rupturing of the posterior part of the annulus, as part of the degenerative process, may take place silently over many years. When the annulus ruptures completely, the nuclear substance escapes. If it is in a semiliquid state, it may be dispersed up and down the canal and may be absorbed. If it is coagulated and contains fragments of hyaline or fibrocartilage, the mass may compress a nerve root and cause pain. Some nuclear material may become inspissated and fibrosed without escaping from the disk.

After extrusion of the disk, although the spine is unstable as shown by subluxation on flexion and extension, narrowing of the disk space requires a considerable period of time. Gradually, the disk space narrows, the vertebrae subluxate further, and reactive sclerosis of the bone adjacent to the disk develops

Rupture of the annulus is followed by invasion of highly vascular connective tissue which tends to heal the defect and absorbs the degenerated tissue.

The cartilage covering the bony end plates is also subject to degeneration with advancing age. It becomes fissured and allows the nuclear material to escape into the adjacent cancellous bone. The latter reacts by encapsulating the herniated tissue with a wall of fibrous tissue and bone. This is the Schmorl's node.

Various Types of Disk Lesions Found at Surgery

1. A soft, fluctuant, thin-walled domelike swelling of the disk. When this is incised, the nuclear material exudes, usually glistening and jellylike in younger patients, containing ragged sequestrae in older patients.

2. A flat or even concave disk, with a soft, thin, bluish or brownish surface tissue, easily punctured with the escape of a small amount of fluid and small amount of sequestrae. If the spine is hyperextended, the disk may bulge more prominently out into the canal.

3. A firm, white, prominent, fixed domelike or irregularly rounded or nodular swelling in the floor of the canal, which is covered by a thin connective-tissue membrane. The root may be stretched over the mass and be reddened and swollen. Occasionally, because of prolonged pressure the nerve root may be thin, flat and bandlike and stretched over and adherent to the disk mass. The covering connective tissue must be excised before rather

[5] Saunders, J B de C M, and *Inman*, V. T: Pathology of the intervertebral disc, Arch. Surg 40.389, 1940.

[6] Hirsch, C, and Schajowicz, F.: Studies on structural changes in the lumbar annulus fibrosus. Acta Orthop. scandinav. 22:184, 1953.

large, gray, necrotic-appearing pieces may be pulled out.

4. Degenerated and sequestrated disk material may lie free within the canal or caught beneath a nerve root.

5. The disk cavity may be obliterated and the disk collapsed. The nuclear material has been lost by inspissation or absorption, and the hole in the annulus has healed. Only a small amount of disk material or scar tissue can be removed by a curette when the annulus is incised.

Other Types of Disk Lesions

In *osteoporosis* affecting the entire spinal column, the trabeculae lessen in size and number, thereby weakening the bony resistance to expansile pressures of the disks. In consequence, the vertebral bodies are indented on their superior and inferior aspects by *normal disks*. These ballooned disks cause the "fish vertebrae" appearance of the spine.

Thinned disks throughout the spine occur in older people because of progressive desiccation and degeneration of the nuclei. When a single disk is narrowed, it probably has resulted from rupture posteriorly, rupture into the adjacent vertebral body (fracture of the plate), or local desiccation.

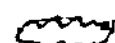
Schmorl's node has been described above. It is a prolapse of nuclear material through a small defect in the cartilaginous plate. A congenital condition which causes larger than normal openings in the bony end plates permits many herniations throughout the spine. These are characterized by their occurring in young individuals, their uniformity of size and shape and by being invariably situated opposite the regions of greatest nuclear expansion. In older people, the herniations are single, larger, irregular. The defects in the cartilage and the end plate is larger. Granulation tissue may enter the disk from the marrow by way of the opening. The prolapsed material may change to cartilage or may ossify. An important result of the herniation is that the disk becomes thinned, the axis of motion is displaced posteriorly, and, as a result of the added strain thrown upon the posterior facets, a traumatic arthritis of the posterior articulations ensues.

Calcification of the disk may occur in the annulus, the cartilaginous plate, the nucleus pulposus, or in the entire disk. The calcified mass is indicative of necrosis; therefore, the disk is subject to rupture posteriorly. As a rule, the calcium deposit absorbs spontaneously after extrusion. (See section on "Calcification of Disks.")

Invasion by infection can occur through the plate from the vertebra, down the longitudinal ligaments, or through the blood stream. Brucellar infection apparently directly destroys the disk. Other infections, as tuberculosis, probably act indirectly by destroying the adjacent vertebrae.

Invasion by a malignant lesion is uncommon. The disk is apparently resistant because of the protective cartilaginous plate.

RUPTURED INTERVERTEBRAL DISK



Etiology

Degenerative changes with advancing age makes the disk increasingly susceptible to trauma. Anything which suddenly or continually increases the superincumbent pressure stresses will eventually cause the posterior fibers of the annulus to give way. Examples of injury include lifting, a fall on the buttocks, direct trauma to the back, twisting movements, or an occupation which involves flexion and lifting motions. The disk may be further weakened under certain circumstances so that the rupture may be effected by a trivial trauma. The disk is notably softened and may rupture during pregnancy and labor and after prolonged bed confinement for an acute illness. When disk rupture occurs without apparent cause, it can be assumed that the degenerative process in itself is sufficient to cause the annulus to yield to mere weight-bearing.

Clinical Picture

The most common history is that of severe low back pain coming on immediately after or within a few hours of an injury. The pain is associated with muscle spasm and flattening of the lumbar area; it is accentuated by forward bending, coughing, sneezing and straining; it is relieved by recumbency. The acute attack subsides within a few days. Many such attacks are the rule, each one

coming on with a lesser amount of trauma, becoming progressively more intense and lasting a longer time, with the interval between attacks being shorter. The course is essentially repeated acute strains of the lumbosacral junction. Sciatic pain eventually develops after the appearance of low back pain. It may appear with the initial attack but ordinarily makes its appearance gradually with subsequent attacks. As the sciatic pain increases in intensity and extent, the backache becomes less and occasionally may be entirely absent. The sciatic pain first appears as an ache in the buttock. This is followed by pain in the posterior thigh area, the popliteal area, the calf or the anterolateral aspect of the leg, and even into the heel, the ankle and the foot. Sciatic pain is most often unilateral and is aggravated by coughing, sneezing and straining at the stool A sensation of muscle cramping in the posterior thigh or calf is a common complaint Intermittency of symptoms is characteristic, and each succeeding attack is usually more severe The following are the main features:

Pain. Sciatic in distribution, accentuated by straining, relieved by recumbency (spinal tumor is often made worse by rest) It is associated with subjective sensations of paresthesias and numbness in about 70 per cent of cases, it is referred to the involved dermatome A lumbosacral lesion causes pain and paresthesias in the sole and the outer border of the foot; an L4-5 lesion is referred to the dorsum of the foot and the big toe; an L2-3 or L3-4 lesion is referred to the anterior thigh, radiating to the anteromedial aspect of the leg.

Reduced Mobility. Forward bending in particular is restricted. In the acute stage with marked muscle spasm, all mobility is restricted.

List of Trunk. The lumbar spine most often deviates away from the affected side, rarely does it lean toward the same side Sometimes the list may alternate, particularly when manipulation is attempted. Any attempt to correct the lateral tilt often increases the pain in the affected limb The disk is usually lateral to the nerve root, and the tilt of the spine away from the affected side may be an attempt to separate the nerve from the protruding tissue. Conversely, protrusion medial to the nerve root, a less common type, causes a list to the painful side. The list is often accentuated when bending forward.

Tenderness. Deep pressure is exerted over the interspace to one side of the mid-line. Either local pain or sciatic pain may be complained of. As a general rule, when pressure or percussion will produce sciatic pain, a large protruded disk will be found.

Nerve Tension Signs.

Flexion of the head and the neck produces cephalad movement of the dural theca and consequent increase in tension on the extradural nerves in the spinal canal.

Flexion of the hip with the knee extended and the ankle dorsiflexed causes caudad movement of the theca and tension on extradural nerves entering the sacral plexus.

Extension of the hip and flexion of the knee causes tension on the femoral nerve roots, L2,3,4.

Jugular compression causes the theca to swell, and the upper attachment of the extradural nerve is carried laterally. As a general rule, if pain is accentuated, the disk protrusion lies lateral to the nerve root.

Tests. The *straight-leg-raising test,* in which the hip is flexed and the knee kept extended, is known as the *Lasègue test.* A positive test is one in which pain in the sciatic area is produced by the maneuver. The *well-leg-raising test* is performed by elevating the extended unaffected leg. Reproduction of sciatic pain on the opposite leg is strong presumptive evidence of a ruptured disk.[7] These ruptures are often medial to the nerve root. When the extended leg is raised to a point just short of producing pain, dorsiflexion of the foot will bring out the sciatic pain. This confirmatory test is known as the *Fajersztajn test.*

Neurologic Signs

MOTOR SIGNS are present in about 96 per cent of cases:

1. *Atrophy* of leg muscles, determined by measuring leg circumferences

2 *Muscle weakness.* Weakness of dorsiflexion of the large toe and inability to walk on the heels indicates 5th lumbar root involvement by the 4th lumbar disk. Inability to

[7] Woodhall, B , and Hayes, G J · The well-leg-raising test of Fajersztajn in the diagnosis of ruptured lumbar intervertebral disc, J. Bone & Joint Surg 32A:786, 1950

walk on the toes because of calf muscle paresis points to 1st sacral root involvement by the 5th lumbar disk.

SENSORY SIGNS are found in 80 per cent of patients.

1. *Fourth lumbar dermatome*, affected by the 3rd lumbar disk, causes a sensory deficit on the anteromedial aspect of the leg and the inner ankle and the foot.

2. *Fifth lumbar dermatome*, affected by the 4th lumbar disk, causes a sensory deficit on the anterolateral aspect of the leg, the medial aspect of the dorsum of the foot, the dorsum of the great toe and occasionally the 2nd toe.

3. *First sacral dermatome*, affected by the 5th lumbar disk, causes a sensory deficit on the posterolateral aspect of the lower leg, the outer foot and the outer 2 or 3 toes.

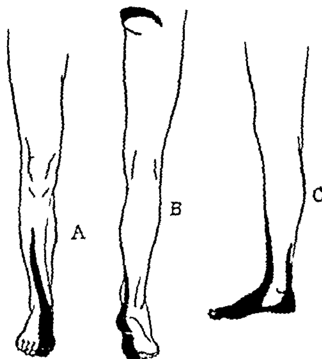


FIG. 474. Sensory loss of 5th lumbar nerve root due to compression at the 4th lumbar interspace.

REFLEX CHANGES

Reflexes of the ankle and the knee are reduced when the 1st sacral and the 4th lumbar nerve root, respectively, are involved. When the deep reflexes are depressed throughout, they may be compared by reinforcement.

ROENTGENOLOGIC FINDINGS

Anteroposterior, lateral and oblique views are taken. A newly ruptured disk is not associated with narrowing of the disk space. Degenerative changes in the posterior articulations attest to concomitant degeneration of the annulus, predisposing to rupture at that level. Subtle indications of disk rupture may

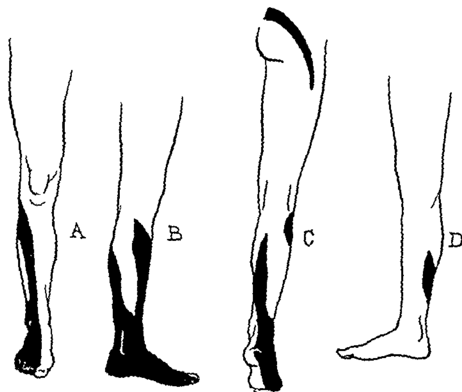


FIG. 475. Sensory loss of 1st sacral nerve root due to compression at the 5th lumbar interspace.



FIG. 476 Pantopaque myelogram. The oblique view more readily displays the degree of defect at the lumbosacral disk level.

be found. The upper vertebra settles downward and necessarily slightly backward as the facets subluxate. Thus, the posterior margins are not aligned. Sacralization of the last lumbar vertebra, if bilateral, excludes this site from suspicion. If the sacralization is unilateral, the lumbosacral disk must be considered. Marked narrowing of the disk space is evidence of an old ruptured or degenerated disk which has become inspissated or fibrosed in situ or after rupture, the narrowing and the bony changes taking place over a number of years. A fresh disk rupture is more likely to originate from another site without bony changes.

MYELOGRAPHY

The diagnosis of a ruptured disk may be confirmed and its position accurately defined prior to surgery by injecting a radiopaque substance into the dural space and taking roentgenograms. *Various types of indentation* defects are noted. A small disk protrusion, particularly in the spacious lower lumbar canal, may fail to indent the dura and may go unrecognized. In addition, the dura may not extend sufficiently distally, and the column of fluid does not reach the level of the disk. Therefore, the failure to demonstrate an indentation defect does not rule out a ruptured disk. On the other hand, a defect, with rare exceptions, indicates a disk protrusion. The indentation defect typically is smooth and unilateral and best revealed by oblique views. Occasionally, a bilateral defect, the hourglass constriction, suggests a mid-line protrusion. A third type of defect is the failure of filling of a nerve root sleeve, particularly when the same root on the opposite side fills and appears to be normal. Striations in the column are seen with large disk ruptures with severe nerve root pressure. These are due to edema of nerve roots. A disk rupture practi*cally never produces a complete block.*

Technic. The patient is premedicated 1 hour previously. He is placed in the hyperflexed position, either sitting or prone, to make the spinous processes more prominent and open up the interspinous intervals. A large 16-gauge spinal needle is used. After sterile preparation and local anesthesia, the needle is inserted in the 3rd lumbar interspace. Manometric readings are taken and changes noted with unilateral and bilateral jugular compression. Spinal fluid specimens are withdrawn for total protein, cell count and other determinations. Six cc. of pantopaque, previously warmed to body temperature, is injected slowly. Too rapid injection of a cold oil will cause it to break up into fine globules, rendering the examination difficult. After proper eye accommodation, the column of pantopaque is followed under the fluoroscope as it flows through the lumbar canal. If any irregularity or other defect is noted, a spot film is made. The defect is visualized better by an oblique view, occasionally by a lateral view. Routinely, anteroposterior and right and left oblique spot views are made at the 3rd, the 4th and the 5th spaces. When a certain area is suspected but *not definitely implicated by fluoroscopy,* a lateral view taken with the patient prone and the back hyperextended will often bring out the indentation defect. At the completion of

the examination, the fluid is centered about the needle and withdrawn. By placing the tip of the needle anteriorly in the canal, much of the pantopaque can be removed. The patient is advised to remain flat in bed for 24 hours to lessen the intensity of headaches. Demerol is very effective in controlling postspinal headache.

Differential Diagnosis

The following are the most important conditions to be considered:

1. **Spondylolisthesis** closely simulates a lumbar disk syndrome. The radiation of pain corresponds to the 5th lumbar nerve root. The typical step defect is palpable in the back. Oblique x-ray views show a defect in the pars interarticularis.

2. **Marie-Strümpell Arthritis.** Limited chest expansion, flattening of the dorsal and lumbar spine, early involvement of the sacroiliac joints, increased sedimentation rate.

3. **Osteoporosis of the Spine.** The patient is either postmenopausal or senile; roentgenograms show demineralization, disk expansion and biconcave vertebral bodies at many levels.

4. **Fracture of a Lumbar Vertebra.** History of severe injury, deformity of the vertebral body. However, a disk rupture is often associated.

5. **Infections of the Spine.** Especially tuberculosis, brucellosis and pyogenic osteomyelitis. Increased sedimentation rate, agglutination tests, skin tests, x-ray findings and constitutional symptoms make the diagnosis.

6. **Primary Tumors of the Vertebra.** Most common are the hemangiomata.

7. **Metastatic Tumors of the Vertebra.** Destruction in several areas are seen in roentgenograms, weight loss, origin often in thyroid, breast and prostate.

8. **Nerve Tumors.** Especially a neurofibroma of the cauda equina. Tumors of the cauda equina typically cause low back pain at first without neurologic signs. The pain radiates to the back or the front of the thigh or the perineum, or it may be sciatic in distribution. Later, symptoms are muscle weakness, flaccid paralysis, and impairment of all forms of sensation. When the lesion occurs low in the spinal canal, sphincter involvement is late. Higher lesions about the conus medullaris have early and severe bladder and rectal changes. Saddle anesthesia is present; ankle jerks are absent; the power of erection and ejaculation in the male is lost; there may be loss of pain and temperature sense. In lesions of the epiconus, paralysis of the feet is an early sign. The following are differentiating signs.[8]

A. Insidious onset of pain without history of trauma

B. Unremitting progressive course

C. Pain constant, unrelieved by recumbency, prevents sleep at night.

D. Myelogram defect typical of tumor

E. Total protein over 100 mg./100 cc. definitely suggests a tumor.

9. **True Sciatic Neuritis.** The nerve is very tender along its entire course. Diabetes, alcoholism, avitaminosis and heavy metals are sought as the cause. The onset occurs without trauma; pain is unrelieved by recumbency; and multiple nerve involvement is usual.

10. **Hip Pathology.** Tumors and arthritis of the hip often cause pain in the buttock and the thigh. Strain thrown on the lower back by hip deformity or a coxalgic gait causes low back pain.

11. **Conditions in the Soft Tissues of the Back.** Any disease process in this area can trigger reflex sciatic pain. Myofascitis and episacral lipoma are common offenders. Infiltration of the suspected area with a local anesthetic relieves the sciatic pain and establishes the diagnosis.

12. **Pelvic Pathology.** The roots of the lumbosacral plexus which make up the sciatic nerve run along the posterior wall of the pelvis before entering the sciatic notch. Infections, e.g., prostatitis, and tumors, e.g., fibroids, easily affect these structures and cause sciatica.

13. **Malformations of Lumbar Spinal Nerve Roots.** Various anomalies are found Two roots may emerge from the common dural pouch and pass through separate foramina; or they may leave the canal through a common foramen. Plexiform connections may exist between nerve roots. These malformations cause pain similar to that caused by a ruptured disk. Decompression relieves the pain.

[8] Toumey, J. W , Poppen, J L , and Hurley, M T.: Cauda equina tumors as a cause of the low back syndrome, J. Bone & Joint Surg. 32A:249, 1950.

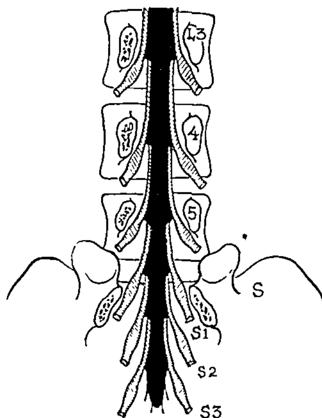


FIG. 477. The relationships of the thecal sac and the lower lumbar and sacral nerves to the pedicles and the intervertebral disks. The subarachnoid space is filled with iodized oil and is represented in black. It is limited laterally by successive nerve roots. The subarachnoid space extends beneath each root as it enters the dural sleeve, forming the axillary pouch The 5th lumbar nerve is contained within the dural sac opposite the 4th lumbar disk, but the 1st sacral nerve enters its dural sleeve above the lumbosacral disk (Redrawn from Bradford, F. K., and Spurling, R. G.. The Intervertebral Disc, ed. 2, Springfield, Ill , Thomas)

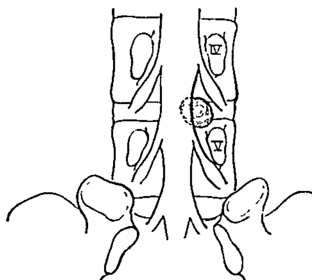


FIG. 478. The usual relationship of the protruded disk at the 4th lumbar interspace. The 5th lumbar nerve root is compressed.

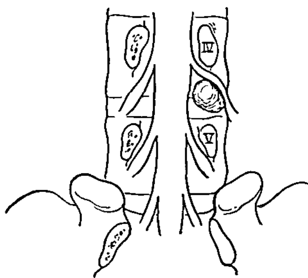


FIG. 479 An uncommon relationship of the protruded disk at the 4th lumbar interspace. The 4th lumbar nerve root is compressed.

14. Varicosities. Symptoms may be initiated by trauma, which probably causes a peridural hemorrhage. Multiple filling defects are noted in the myelogram.

15. Vascular Insufficiency. Reduction of blood flow to an extremity may be sudden (embolus) or gradually progressive (arteriosclerotic narrowing of the vascular lumen) and causes ischemic pain The arterial occlusion may be incomplete and partially compensated by the development of a collateral circulation so that pain in the extremity occurs only with activity (intermittent claudication). When vascular obstruction is more nearly complete and the blood flow is unable to meet the needs of the muscle and nerve structures under any circumstances, rest pain develops. By holding the extremity in the dependent position, the patient utilizes the force of gravity to increase blood flow to the distal areas and to reduce rest pain.

Ischemic pain, whether intermittent claudication or the severe rest pain, appears in the calf if the occlusion is in the femoral artery;

in the hip if the obstruction is in the iliac artery; or in both hips or the lower back if the occlusion is in the aorta. Arteriography is diagnostic.

Treatment

Protruded disk tissue can be resorbed by granulation tissue and the defect in the annulus repaired by fibrous tissue.[9] Removal of pressure from the nerve root relieves the symptoms. However, fibrosis may engulf the dura and the nerve root and cause persistent low-grade symptoms. As a rule, about 90 per cent of initial attacks subside with conservative treatment. The healed area of the posterior annulus is thinner, more fragile and less resistant to pressure. Therefore, each subsequent attack becomes less responsive to the natural healing process. However, repeated small protrusions and dissolution of a small portion of the disk each time eventually may lead to complete loss of the disk and a permanent cure.

Conservative treatment is indicated for each acute attack. This attitude must be modified by various factors, such as the economic hardship imposed by repeated hospitalization, intolerance to pain requiring the use of narcotics, important neurologic loss, etc. The only absolute indication for operation is persistence of symptoms in spite of conservatism, or if attacks are so frequent as to constitute one continuous unyielding attack.

Conservative Treatment. Rest in bed is important in removing the weight-bearing pressure from the disk. The position is one in which the hips and the knees are flexed, thereby relaxing the sciatic nerve and reducing the lumbar lordotic curve which opens the posterior disk intervals. Traction of 15 pounds is applied to both legs or through a pelvic traction band, the body above the pelvis acting as countertraction by elevating the foot of the bed. This theoretically distracts the disk interval and permits replacement of the protruded tissue. A firm mattress and a fracture board may give some comfort. Heat is applied to the lower back by an electric pad, an infrared lamp, hot towels, or diathermy. It may be followed by light massage.

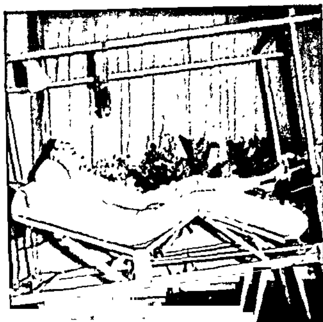


Fig. 480. Bilateral leg traction for low back condition. Hip and knee are flexed, reducing the lumbar lordosis, countertraction obtained by elevating the foot of the bed.

Sedatives are given to reduce muscle spasm.

Some orthopaedic surgeons practice manipulation in an effort at redisplacing the disk. This treatment is regarded as controversial and a form of quackery by many men. However, the author has attempted the maneuver in patients who did not respond to bed rest and were regarded as candidates for surgery. Occasionally, the result was dramatic.

The method is performed as follows:[10] The patient lies on his side on the edge of the table facing the surgeon, and the leg that is up is allowed to drop forward over the edge of the table, carrying forward that side of the pelvis. The arm that is up is placed backward behind the patient, pulling the shoulder back. Then the surgeon places one hand on the shoulder and the other on the iliac crest and twists the torso by pushing the shoulder backward and the iliac crest forward. The maneuver is sudden and forceful and frequently is associated with an audible and palpable crunching sound in the lower back. When this is felt, the relief of pain is usually immediate. The maneuver is repeated with the patient on the opposite side. Finally, the pa-

[9] Lindblom, K., and Hultquist, G. Absorption of disc tissue. J. Bone & Joint Surg 32A.557, 1950

[10] Ramsey, R H Conservative treatment of disc lesions, Am. Acad Orthop Surgeons, Lect vol. XI, 1954.

tient is turned on his back, and the hips and the knees are hyperflexed sufficiently to flex the lumbar spine forcibly; this tends to open up the disk spaces posteriorly. The patient should be cautioned beforehand that the manipulation may make his symptoms worse and that this is an attempt to avoid surgery.

The period of bed rest varies from several days to a few weeks before the acute symptoms subside. A low back support is fitted before the patient is allowed to be ambulatory. This consists of a corset, a brace or a plaster cast. The cast should be of the flexion jacket type. A corset or a brace permits removal when the patient is recumbent. The immobilization should extend from the dorsolumbar junction above to the greater trochanters below. Because restricted back motion progressively weakens the musculature and causes further degeneration of spinal structures, the back support is discarded as soon as symptoms have been relieved. Exercises are prescribed to strengthen the back extensors, the gluteus maximus, the abdominals and the quadriceps, all of which assist the lifting movement. During this period of rehabilitation, lifting and bending are forbidden. Short rest periods are advisable. Proper posture is practiced Weight reduction is effected by diet and medication. When the patient has been regarded as fully recovered, certain athletic activities are curtailed. Bowling and handball are notable instigators of low back conditions. When a slight recurrence of low back pain or sciatica is noticed, it should be regarded with alarm The back support is reapplied, and frequent periods of rest are in order.

In many cases, this program is sufficient to control symptoms for many years, if not indefinitely. The patient must be made aware of the fact that at any time he may become a candidate for surgery. In spite of successfully surmounting the repeated attacks associated with progressive disk disintegration, localized instability and degenerative arthritis may cause disabling symptoms in later years, necessitating a spine fusion

Surgical Treatment. When symptoms are persistent, the disk must be removed. Loss of the disk throws abnormally increased stresses on the posterior articulations with eventual development of a traumatic degenerative arthritis. This latter condition is capable of causing disabling symptoms, particularly if the patient indulges in a heavy-lifting occupation. The author believes that a spinal fusion should be done at the same time. A controversy has arisen concerning the necessity of spinal fusion. Opponents, especially neurosurgeons, state that the added procedure carries added risk and prolongs the convalescence. Further, they state that the small number of patients who develop later disability can be operated upon at a later date. However, the results of the combined operation are infinitely better than removal of the disk alone.

TECHNIC.[11] Endotracheal anesthesia is preferred, although spinal and local anesthesia may be used. *The patient is placed in the* prone position on the operating table. A large blanket roll is placed on either side of the patient, extending from the shoulder to the groin. This relieves the abdomen from compression, thereby preventing engorgement of the predural venous plexus. The supraspinous ligament and the periosteum are incised over the tips of the 4th and the 5th lumbar and the 1st sacral spinous processes. The lateral muscle masses are elevated from the vertebrae, and hemostasis is secured by packing. Self-retaining retractors are inserted. The interspinous ligaments are removed. The laminae and the spinous processes are curetted clean as far laterally as the facets. The interspace between the laminae of the 5th lumbar and the 1st sacral is wide; that between the 4th and the 5th lumbar laminae is narrow. In the space selected the ligamentum flavum is incised in the mid-line, and a blunt flat dissector is inserted beneath the ligament in a lateral direction. This protects the underlying structures from damage as the ligamentum flavum is incised and removed. At the 4th lumbar interspace, added exposure requires partial removal of the laminae with the Kerrison rongeur. The interspace is further widened by flexing the patient on the table and by inserting a spinous process spreader. The dura and the nerve root are freed of adhesions by blunt dissection and retracted medially by a nerve root retractor. The 5th lumbar root, which is compressed by a disk at the 4th interspace, leaves the dura at a level just below the disk

[11] Bradford, F K, and Spurling, R. G.: The Intervertebral Disc, ed 2, Springfield, Ill, Thomas, 1945

level. The 1st sacral root, which is compressed by the 5th lumbar disk, lies within its own sheath lateral to the dura. Bleeding from the large plexus of veins anterior and lateral to the dural sac may be troublesome. They must be carefully avoided by deliberate blunt dissection. Bleeding is controlled by packing with fibrin foam dipped in thrombin solution and covered with a moist cotton pledget. Gentle suction during the dissection is necessary to detect the site of bleeding.

The herniated mass of nucleus pulposus may lie free within the canal, displacing the nerve root forward or to the inner or outer side. In other instances the capsule may be intact over the mass, and a bulge presents itself. The bulge can be made more prominent by flexing or hyperextending the patient. The loose material is lifted out after incising the covering capsule or scar tissue. A pituitary rongeur is inserted in the opening in the annulus, and the interior of the disk is cleaned out. If the herniation is in the mid-line, bilateral exposure is necessary for complete removal. If the mass is lateral, additional lamina is removed laterally with the Kerrison punch.

The nerve root occasionally will be found to have been constricted by scar tissue and badly damaged. All cicatrix should be carefully removed. If necessary, the intervertebral foramen should be decompressed by removing the overlying facets. The possibility of postoperative pain from nerve damage should be borne in mind. Such an eventuality makes it worthwhile to perform a posterior rhizotomy. The dorsal surface of the dural sleeve is incised along the axis of the nerve root. The dural margins are gently pushed back to expose the glistening posterior root which is separated and sectioned, proximal to the spinal ganglion. Postoperatively, only a hypesthesia or a small area of anesthesia can be expected.

Finally, spinal fusion is performed by any of the available methods. The tissues laterally can be elevated in the plane between the superficial and the deep fascia to reach the crest of the ilium and cancellous and cortical bone grafts removed. The wound is closed in layers. Postoperatively, after a period of recumbency of several weeks, a brace or a cast is applied, and ambulation is permitted.

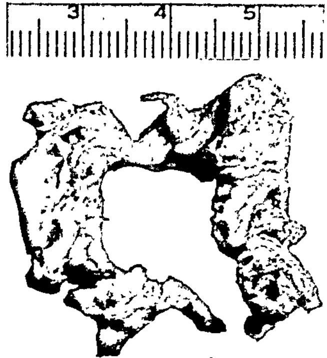


FIG. 481. Example of intervertebral disk material removed at surgery.

RECURRENT SYMPTOMS AFTER DISK REMOVAL.[12] In spite of removal of the disk and the performance of a spinal fusion, symptoms may persist or may recur after a latent period. If immediately after operation the original symptoms and findings continue unabated, the causes include: (1) failure to remove the offending disk, (2) an overlooked second ruptured disk, (3) surgical trauma to the nerve root during removal of the disk, and (4) the presence of other pathology, such as an unrecognized tumor.

Often symptoms are relieved only to recur after a variable period. The causes include: (1) insufficient removal of disk material and further extrusion, (2) rupture of another disk, (3) adhesions about the nerve root and (4) formation of an osteophyte at the site of removal of bone.

Treatment.[12] The bony mass of the previous fusion must be removed, and the spinal canal explored. This requires meticulous excision of all cicatrix and freeing of all neural structures well above the previously operated area. Removal of scar tissue is facilitated by starting at the first normal vertebra above, excising the lamina and working downward. Good hemostasis must be secured to reduce

[12] Turek, S. L.: Failure of disc operations, Am. J. Surg. 87:241, 1954.

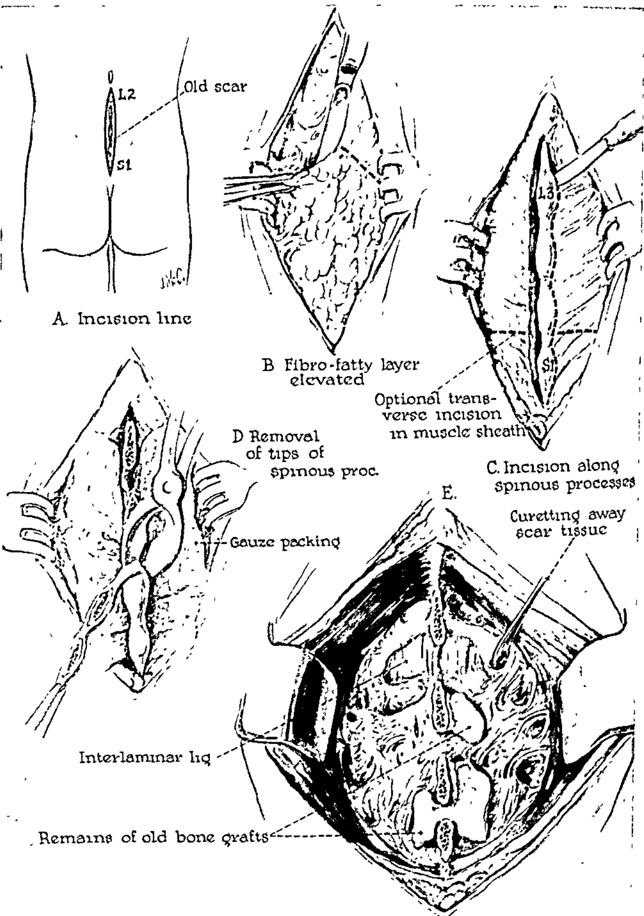
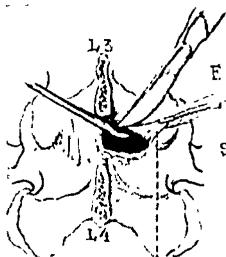
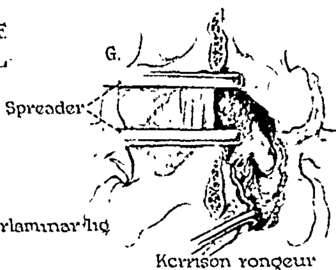
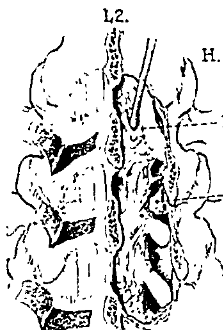
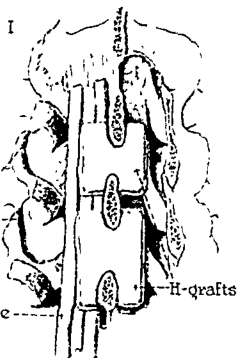


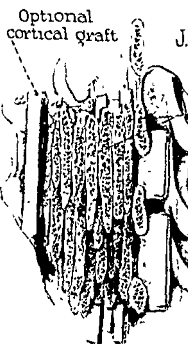
FIG. 482. Reoperation for recurrent symptoms after disk removal. The illustrations depict the necessary steps of exposure and fusion of spine. (Turek, S. L.: Failure of disc operations, Am. J. Surg. 87:241) (*Continued on facing page*)

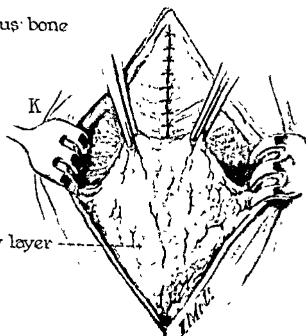

L3
F
L4
Elevation of interlaminar lig


G.
Spreader
Kerrison rongeur


L2.
H.
Fibrous tissue
Exostosis
S1.


I
Cartilage and cortex removed
H-grafts
Cortico-cancellous iliac bone


Optional cortical graft
J.
Cancellous bone strips


K
Fibro-fatty layer

the possibility of scar-tissue formation. The laminae of the contralateral side must be preserved if possible to provide a site for arthrodesis. If extensive removal of the posterior arches is necessary, spinal fusion can be accomplished by laying bone grafts from the sacrum along the transverse processes adjacent to the pedicles. A strong cortical distracting "H" graft may be placed between the sacral spinous process and the spinous process of the first normal vertebra above.

Postoperatively, immobilization by a spica cast extending from the nipple line above to the knee on one side is necessary. To reduce the possibility of adhesions about the nerve roots, straight leg exercises of the nonimmobilized extremity are performed.

WARNING REGARDING SPINAL CORD TUMORS

Tumors of the spinal cord, particularly those about the conus and the cauda equina, can cause symptoms and findings closely simulating an intervertebral disk rupture. The onset may even be acute and the course often intermittent. Failure to find a protruded disk at surgery should suggest exploration of the upper lumbar area, both intradurally and extradurally. The study of this subject on intervertebral disk rupture should be complemented by studying the section on spinal cord tumors (q.v.).

LUMBAR NERVE ROOT COMPRESSION BY AN EXOSTOSIS[13, 14]

A bony prominence may develop within the vertebral canal where it can irritate and compress a nerve root, simulating the clinical picture of disk protrusion.

Etiology

Trauma is the most common cause. Often a history of a fall upon the buttocks is obtained. Surgical trauma in the course of performing a laminectomy may be followed by hyperplastic bone formation.

Degenerative changes rarely produce a spur at the posterior margin of the vertebral body or at the anterior aspect of a facet.

Developmental Cause. In the absence of traumatic or degenerative factors, a developmental cause is possible.

Pathology

The spur often is found at the isthmus projecting anteriorly and medially into the vertebral canal where it compresses the lateral aspect of the dura. The associated articular facet is usually bulbous, irregular and obliquely disposed. Thickening of the ligamentum flavum is often observed at the same level. The most common site for spur formation is at the L4-5 interspace, the point of greatest mobility of the lumbar spine. Therefore, it compresses the 5th lumbar nerve root which still lies intradurally at this level. The foramen above the spur and immediately below the pedicle is not narrowed; therefore, the 4th lumbar nerve root is not compromised. However, when the spur is quite large and lies more proximal, as uncommonly arises from surgical trauma or anteriorly from a degenerative process of the margin of the vertebral body, the nerve root within the foramen may be compressed.

Clinical Picture

Most often a history is obtained of a fall upon the buttocks, followed immediately by low back pain and sciatica.

This suggests a periosteal tear and hemorrhage, causing nerve root irritation. The leg pains are worse than the back pain and are accompanied by numbness and paresthesias. Coughing, sneezing and straining intensify the symptoms, and the picture is almost identical with that of a rupture disk. However, the symptoms and the findings persist in spite of supports and bed rest. Hypesthesia most often is found at the L5 dermatome, the anterior tibial surface, the dorsum of the foot proximal to the great toe and the dorsum of the great toe. Less commonly, involvement of the L4 and the S1 dermatomes is found, indicating spur formation at the L3-4 and the L5-S1 levels, respectively.

[13] Epstein, J. A., and Davidoff, L. M. Recognition and management of spinal cord and nerve root compression caused by osteophytes, Bull Rheumat Dis 3.47, 1953

[14] Schnitker, M. T., and Curtzwiler, F. C. Hypertrophic osteosclerosis (bony spur) of the lumbar spine, J. Neurosurg. 15:121, 1957.



FIG. 483. Anatomic specimen, showing sagittal section of the lumbosacral spine. Note that the nerve root lies in a foramen well above the tip of the inferior facet but can be compressed as subluxation occurs. (Specimen from Dr. Yglesias. Williams, P. C.: Conservative management of lesions of the lumbosacral spine, Am. Acad. Orthop. Surgeons, Lect. 10.90-121)

Roentgenologic Findings

The roentgenogram is usually negative. An enlarged, bulbous facet suggests the location of the lesion. Myelographic study reveals a long concave defect only when the spur is large.

Treatment

Laminectomy is performed, the spur is usually found at the isthmus and removed, and hypertrophied ligamentum flavum is resected. Spine fusion is indicated when degenerative changes affect the facets.

LUMBOSACRAL STRAIN
(The Facet Syndrome[15])

The true joints of the spine are the apophyseal joints formed by the superior and the inferior articular processes of the poste-

[15] Williams, P. C.: Lesions of the lumbosacral spine, Am. Acad. Orthop. Surgeons, Lect., vol. 4, 1947.

rior neural arch. During flexion, extension and rotation of the spine, gliding motions take place between the apposed flat facets, which are covered with hyaline cartilage. The articulation is surrounded by a ligamentous capsular structure which is loose enough to permit motion but becomes taut at the extremes of each movement. Like other ligaments it is subject to *stretching and tearing, particularly by hyperextension,* as a result of which the facets subluxate upon each other.

Predisposing factors which encourage tearing of the capsule and subluxation are: (1) *excessive superimposed weight*; therefore, the lumbosacral articulations are prone; (2) *an acute lumbosacral angle* which aggravates the hyperextension; the normal lumbosacral angle is 120°, an acute lumbosacral angle is caused by forward tilting of the pelvis caused by a taut tensor fascia femoris, an inadequate gluteus maximus, wearing high heels, an extra lumbar vertebra, compensatory lumbar lordosis as a result of increased dorsal kyphosis (vertebral epiphysitis), (3) *degenerative changes of advancing age*; (4) *loss of the intervertebral disk*; (5) *chronic occupational strains* requiring excessive bending and lifting; (6) *vertical disposition of the articular facets.*

When the facets subluxate, the inferior facet is displaced upward and impinges upon the inferior vertebral notch of the upper vertebral body, where it may compress the nerve root. The upper vertebral body settles slightly downward and backward, its inferior margin lying slightly posterior to the upper margin of the lower vertebral body. The posterior portion of the disk space is narrowed. The superior facet glides downward. The intervertebral foramen becomes narrowed. One must remember that the exiting nerve root lies in the upper half of the foramen above the site of the normal articulation and cannot be compromised unless the inferior facet can move upward and compress it.

The superior sacral facets by their impingement against the 5th lumbar vertebral body become weight-bearing structures. As a result, the facets become sclerosed, and the same changes are observed in the notches. The facet articulations themselves develop degenerative changes, including narrowing of the joint space, loss of articular cartilage, sclerosis, irregularity and osteophyte formation These alterations take place over a number of years.

At the outset, tearing of ligaments and subluxation are manifest by local symptoms of low back pain accentuated by the motion which stretches the ligaments, namely, hyperextension. Later, as the nerve root is compressed, sciatica and neurologic findings referable to the 5th lumbar nerve root become prominent. Eventually, symptoms of localized degenerative arthritis are superimposed.

Displacement of the facets occasionally throws sufficient strain upon the intervertebral disk to rupture the latter. However, most commonly protrusion of the disk accompanies a flexion injury.

Clinical Picture

A history is often given of repeated episodes of acute low back pain caused by forceful hyperextension. Between attacks, hyperextension movements of the back increases the pain whereas flexion reduces it. Examples of hyperextension include sleeping on the abdomen, sitting in an erect attitude, bending over a wash basin, lifting a load in front of the body at or above the waist line, working with the hands and the arms above the head, and arising from a sitting position. When the symptoms are acute, sneezing and coughing accentuate the pain unless done with the knees and the chest approximated. As time goes on symptoms of nerve root irritation appear, including sciatica, paresthesias, numbness over the dorsomedial aspect of the foot, and weakness of dorsiflexion of the large toe. When localized degenerative arthritis has developed, complaints are pain and stiffness of the lower back after rest and with changes of weather, relieved by heat.

On examination, the patient stands with a flexion attitude to relieve the ligamentous strain. Localized tenderness is observed about the lumbosacral junction. In the acute stage the back muscles are in spasm, and motions are restricted in all directions. In the relatively pain-free interval, backward bending is prevented by the increase of discomfort. An increased lumbar lordosis and any rounding of the dorsal spine should be noted. The



Fig. 484. Lateral view of the lumbosacral spine, showing posterior displacement of the 5th lumbar vertebra and lodging of the sacral facets in the inferior notches of the 5th lumbar. Rounding of the superior margins of the sacral facets is of degenerative origin. (Williams, P. C.: Conservative management of lesions of the lumbosacral spine, Am Acad. Orthop. Surgeons, Lect. 10 90-121)



Fig. 485. Oblique view of the lumbosacral spine, showing complete subluxation of the lumbosacral facets. (Williams, P C.: Conservative management of lesions of the lumbosacral spine, Am. Acad. Orthop. Surgeons, Lect. 10:90-121)

straight-leg-raising test may accentuate the low back pain at the extreme point of the maneuver, because the capsular structures are stretched. In addition to observing forward tilting of the pelvis, the usual causes of this condition are sought.

By the Ober test tautness of the fascia lata can be determined This is performed by placing the patient on the unaffected side, flexing the affected hip, then widely abducting it, extending the hip, and finally allowing the thigh to drop toward the adducted position. When the tensor is tight, the thigh will be sustained momentarily in the abducted position before falling toward the table. Flexion contracture of the hip is determined by the Thomas test. Inequality of the lower limbs will cause a lumbosacral strain. The lengths must be recorded

When symptoms of nerve root irritation are present, a neurologic examination is performed. Loss of sensation over the dorsomedial aspect of the foot, weakness of dorsiflexion of the large toe, and intactness of the ankle reflex point to 5th lumbar nerve root involvement. In the presence of tenderness localized to the lumbosacral junction and positive x-ray findings, a protruded disk cannot be implicated at this level. Instead, a mechanical lesion is the usual cause.

Roentgenologic Findings

Oblique studies of the lumbosacral area are necessary to display the facets. This reveals subluxation of the facets, posterior displacement of the segment immediately above, some narrowing of the posterior portion of the disk space, lodging of the superior margins of the

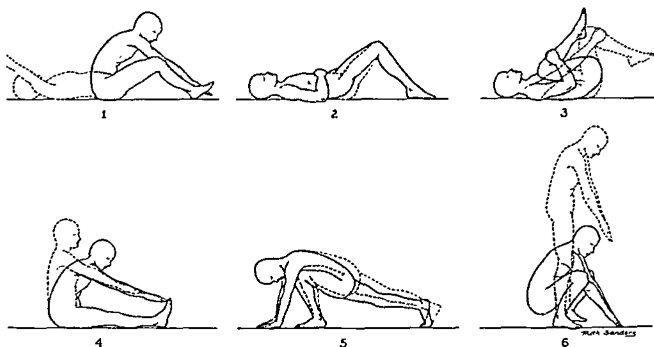


Fig. 486. Postural exercises designed to reduce the lumbosacral angle. (Williams, P. C.: Conservative management of lesions of the lumbosacral spine, Am. Acad. Orthop. Surgeons, Lect. 10:90-121)

sacral facets in the inferior vertebral notches of the 5th lumbar vertebra and, at a late date, degenerative arthritic changes localized to the articulations at this level. The intervertebral foramina at the lumbosacral junction are narrowed This finding is generally extreme when nerve root symptoms are present.

Treatment

This is directed toward reducing the hyperextension and therefore the subluxation. Improvement in the strength of the abdominals and the gluteus maximus muscles promotes flexion at the lumbosacral junction and restores the sacrum to the more vertical position. The hip flexors must not be strengthened, because they tend to bend the pelvis forward and to make the lumbosacral angle more acute. Conversely, flexion contracture of the hip must be overcome.

Chronic Stage. The patient is instructed on avoiding harmful attitudes, such as sleeping in the prone position, rising from a sitting position, bending over a wash basin, etc. Arduous occupational activities are prohibited. A heel lift corrects lower limb inequality

Exercises are aimed at developing the flexor muscles of the lumbosacral spine These are done on a padded floor several times daily Exercise 1 (Fig. 486) develops the abdominal muscles. The hip flexors must be relaxed so that the hips and the knees must be flexed, and the feet must not be anchored. Exercise 2 (Fig. 486) develops the gluteus maximi. The pelvis is rotated forward. The hands are placed on the abdomen above the umbilicus so that flexion takes place only at the lumbosacral spine. Exercise 3 (Fig. 486) passively stretches the erector spinae and the contracted fascia and ligaments over the posterior aspect of the lumbosacral junction. The thighs are spread apart, and the knees are pulled back and forth toward the axillae. Exercise 4 (Fig. 486) not only stretches the erector spinae and the posterior fascial structures but also the hamstrings. This movement obviously cannot be used in the presence of nerve root involvement. Exercise 5 (Fig. 486) stretches the structures of the anterior thigh which limit extension of the hip joint. These consist primarily of the fascia lata and the iliofemoral ligament. Exercise 6 (Fig. 486), the "flat footed squat," strongly flexes the lumbosacral junction. It strengthens the quadriceps and by repeated drilling develops the habit pattern of bending with the knees flexed rather than bending at the hips alone. The feet are placed flat upon the floor about 12 inches

apart, and the patient squats up and down. The heels must not be raised from the floor.

The forward bent attitude must be encouraged and practiced. The body weight should be borne on the heels. When carrying the weight on the balls of the feet, such as when wearing high heels, the pelvis is thrust forward with a compensatory backward thrust of the upper spine and a resultant increase of lumbosacral extension. A load should be lifted with the legs and the lumbosacral spine in flexion and not carried in front of the body above the waist line. Sitting is done with the buttocks "tucked under." Too high a chair aggravates lumbosacral extension. In driving, the seat should be pushed forward to raise the knees and obliterate the lumbar lordosis. Sleeping on the abdomen is avoided. Sleeping on the back with the knees elevated is recommended. When lying on the side, the knees and the hips are flexed. A soft mattress will permit lumbosacral extension. A firm mattress or a board under the mattress encourages the lumbar spine to sag backward with reduction of discomfort.

A Williams hollow back brace is worn when the patient is ambulatory. In principle, this effects 3-point pressure to reduce lumbosacral extension. Posteriorly, a transversely placed bar at the lower thoracic area and another over the buttocks effect forward pressure, while anteriorly an abdominal band exerts backward pressure.

A flexion plaster-of-Paris jacket utilizes

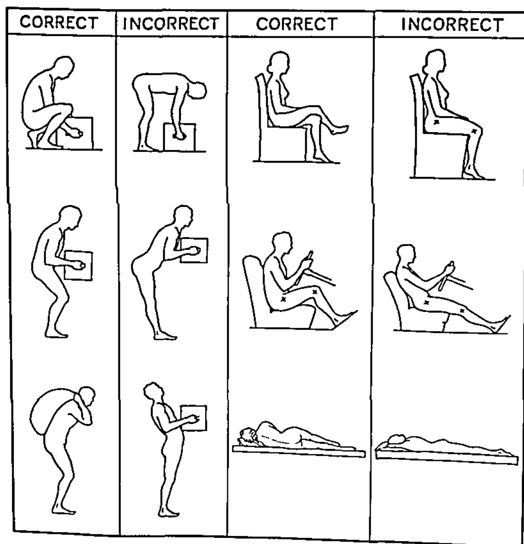


FIG 487. Correct and incorrect postural attitudes. (Williams, P. C.: Conservative management of lesions of the lumbosacral spine, Am. Acad. Orthop. Surgeons, Lect. 10:90-121)



FIG. 488. The Williams lumbosacral flexion brace (hollow back brace). (Williams, P. C.: Conservative management of lesions of the lumbosacral spine, Am. Acad. Orthop. Surgeons, Lect 10:90-121)

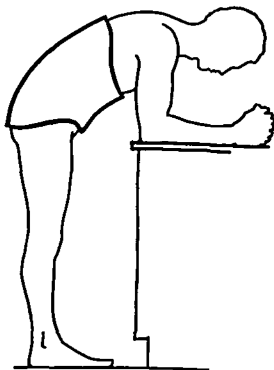


FIG. 489. Flexion plaster-of-Paris jacket. (Williams, P. C: Conservative management of lesions of the lumbosacral spine, Am. Acad Orthop. Surgeons, Lect 10:90-121)

the same principle and immobilizes the back more effectively. However, it should not be *used until full flexion has been restored to* the lumbosacral spine. The patient stands in the forward bent position with the elbows resting on a table. This reduces the acuteness of the lumbosacral angle and lessens the subluxation of the articulations. The cast is applied so that the back is enclosed from the lower thorax above to the lower buttocks below. In front, the cast extends from the *xiphoid to the symphysis pubis. As the* patient straightens up, he experiences a sensation of pressure over the abdomen anteriorly and at the upper and the lower edges of the cast posteriorly. The cast may be cut vertically in front for removal and drying, then reapplied with encircling bands. The patient is instructed to wear the cast only when ambulatory, removing it only for exercises. *Discontinuance of the back support is permissible* only when symptoms have subsided, but exercising must be continued indefinitely.

Acute Stage. Absolute bed rest in the flexed position is required. The back rest and the knee rest are elevated. Pelvic or bilateral leg traction with 30 pounds of weights is applied, and the foot of the bed is elevated for counter-

traction. When the pain has reduced in intensity, Exercises 1 and 2 are started and are increased gradually each day. Heat is applied to the lower back, and sedatives are administered. Finally, a flexion jacket is applied, and the patient is allowed to be ambulatory as exercises are continued.

If symptoms persist for more than 2 or 3 weeks, it may be advisable to explore the spine preceded by myelographic study. A protruded intervertebral disk may be present even in the absence of a myelographic defect. Lumbosacral fusion is necessary. At the same time the nerve roots must be decompressed by removing the articular processes.

Occasionally, a marked lordosis and flexion contracture of the hip may be defined as due primarily to a tight iliotibial band. Section of the band and the adjacent portion of the intermuscular septum is indicated.

SPRUNG BACK

"Sprung back" is the term applied by Newman[16] to a condition of low back pain caused by tearing of all the posterior supportive ligaments of the lumbosacral junction, particularly the supraspinous ligament, and manifest clinically by pain in the *flexed* position. In contrast, the facet syndrome is typified by pain in the hyperextended position.

Clinical Picture

The patient is often a female between 15 and 35 years of age. The discomfort complained of is a dull, nagging ache which is worse in positions of flexion of the lumbar spine. Examples are: sitting in a slouched position with the lower back unsupported, and relieved by sitting straight with a pillow in the small of the back, lifting a weight at arm's length; bending and stooping with the knees fully extended.

On examination, lumbar lordosis is prominent Muscle spasm, when present, is mild. Active flexion of the spine is normal. The straight-leg-raising test may cause some low back pain but is not restricted as contrasted with nerve root compression The nerve roots are uninvolved; consequently, the neurologic examination is negative. To determine whether or not the supraspinous ligament is at fault, the patient is placed in the prone position with 3 or 4 pillows beneath the abdomen to provide relaxation in the semiflexed position. This opens up the gap between the spinous processes and facilitates palpation of the supraspinous ligaments. Tenderness is found in the mid-line between spinous processes L5-S1 or L4-L5. A depression denotes damage or laxity of the supraspinous ligament and contrasts markedly with the resistance of the intact ligament perceived at other levels. Small, firm nodules can often be palpated at the site of tenderness. If hypertonic saline is injected into the suspected ligament, the local and referred pain will be intensified if that ligament is at fault. On the other hand, injection of a local anesthetic relieves the pain.

The x-ray examination is usually negative.

Mechanism of Injury

The multifidus muscles attach to the tips of the spinous processes and act as a braking mechanism while the spine is flexed. The erector spinae muscles extend the spine as a whole. The lumbar fascia meets in a raphe in the mid-line where it attaches as the supraspinous ligament to the tips of the spinous processes; this acts to check overflexion of the spine. The supraspinous ligament is strong. The interspinous ligaments and the joint capsules are less strong but can be torn only after the supraspinous ligament tears.

Overflexion of the lower lumbar spine produces excessive posterior strain at this level, especially when tight, contracted hamstrings restrict forward movement of the pelvis. The supraspinous ligament is torn. Further flexion causes tearing of the interspinous ligament and the capsule. An extreme degree of flexion will cause further tearing of the posterior longitudinal ligament, thereby weakening this structure for disk prolapse. (Rarely, a compression fracture of the body occurs at the same time when flexion is extreme.) Interruption of the ligaments results in a permanently weak and painful back. Injury about the lumbosacral level is more likely if posterior structures are congenitally weakened, such as when a small sacral spinous process or spina bifida occulta is present. When the 5th lumbar vertebra is sacralized, it is more stable, and the flexion strain is transferred to

[16] Newman, P H · Sprung back, J Bone & Joint Surg 34B 30, 1952.

the L4-L5 level. Occasionally, the capsular ligaments are strong, and the bony structures give way. Fracture of the pars interarticularis results in a spondylolysis. A facet may fracture, or a spinous process may split longitudinally.

The supraspinous and the interspinous ligaments may heal in an elongated position, and the joint capsules may remain lax. Affected vertebrae become unstable, and the joints may subluxate The healing scar tissue is painful when subjected to strain as in the flexed position. Therefore, the patient assumes the extended position to relieve the strain. These frequently recurring strains antedate actual disk rupture, which is caused by a giving-way of the damaged posterior longitudinal ligament. Thereafter, the patient assumes a position of flexion to reduce further extrusion of the disk.

The referral of pain from the L4-L5 level is to the buttocks and the outer anterior thighs. That from the L5-S1 level is to the buttocks, the outer thighs, the calves and the feet.

Treatment

Mild cases are treated similarly to conservative therapy advised for disk cases Sometimes repeated needling of the supraspinous and the interspinous ligaments with a local anesthetic effects relief. Obstinate cases require stabilization by spinal fusion.

THORACIC INTERVERTEBRAL DISK HERNIATION[17]

Clinical Picture

When a disk ruptures in the thoracic region, the nerve root is irritated and compressed early in the course of the disease. Involvement of the spinal cord occurs late. Radicular pain is caused by encroachment by disk tissue on the intervertebral foramen. The discomfort is described as crushing, sharp, burning, or pushing and is accentuated by movements of the spine. The site of disk rupture determines the distribution of referred pain. In the upper thoracic spine, the pain is girdle-like and extends along an intercostal space. When the lower thoracic spine is involved, pain is referred to the abdomen and the groin. Hyperesthesia and hypoesthesia of the affected dermatome localizes the lesion. A superficial abdominal reflex may be reduced.

The majority of thoracic protrusions occur in the region of the 6th to the 11th thoracic disks.[18, 19, 20] It is most common in middle and later life and apparently seems to be related to a degenerative process.

The position of the protrusion determines the clinical picture. A mid-line protrusion causes inconstant pain, pyramidal tract signs, bowel and bladder symptoms and sensory defects. A lateral protrusion causes more pain, radicular in distribution, and fewer cord signs.

Involvement of the 1st thoracic root projects pain down the inner aspect of the arm to the little finger. The sensory loss in the little and half of the ring finger, the ulnar side of the hand and the forearm may be objective or only subjective. The intrinsic muscles of the hand may be weak. Occasionally, a Horner's syndrome may be noted.

The 2nd thoracic root involvement produces radicular pain in the axilla and along the inner aspect of the arm.

Protrusion of the other thoracic disks causes intercostal pain and, when the lower thoracic nerve roots are involved, pain referred to the abdomen or the groin.

Diagnosis

Pertinent diagnostic signs include: (1) pain and tenderness to percussion over the involved disk; (2) narrowing and calcification are often associated; (3) myelography; (4) subarachnoid block and increased protein content of spinal fluid. The absence of a block or increased protein does not rule out a ruptured disk. Conversely, their presence is often associated with a neoplastic or inflammatory lesion.

[17] Epstein, J. A · The syndrome of herniation of the lower thoracic intervertebral discs with nerve root and spinal cord compression, J Neurosurg. 11.525-538, 1954

[18] Love, J. G, and Kiefer, E. J · Root pain and paraplegia due to protrusions of thoracic intervertebral discs, J Neurosurg 7-62, 1950.

[19] Kroll, F W., and Reiss, E.: Der thorakale Bandscheibenprolaps, Deutsche med Wchnschr. 76: 600, 1951

[20] Muller, R · Protrusions of thoracic intervertebral disks with compression of the spinal cord, Acta med scandinav 139 99, 1951.

Myelography

X-ray examination early is negative. Later, an osteophyte narrowing the intervertebral space is seen on oblique views. Early, the spinal fluid is unaltered. Later, the protein is elevated. Jugular compression rarely reveals a block, because the lesion is usually lateral. A myelogram may reveal a partial or a complete block; but the flow of pantopaque through the thoracic region is generally too rapid for roentgenography so that observation under the fluoroscope offers more information

Treatment

When the protrusion is far lateral, operation is not urgent but may be done to relieve pain. Decompression by hemilaminectomy or by removing the roof of the foramen (foraminotomy) is generally adequate. Dorsal rhizotomy ensures relief of pain. The denticulate ligaments are divided to lessen traction on the cord and to permit greater freedom of movement. When the spinal cord is involved, immediate surgery and decompression are mandatory. It is not necessary to remove a bony spur; decompression is sufficient. Prognosis is poor if laminectomy is delayed.

CALCIFICATION OF INTERVERTEBRAL DISKS[21, 22]

Calcification in an intervertebral disk may occur in 3 places: (1) peripherally in the an-

[21] Calvé, J., and Galland, M : Sur Une Affection Particulière de la Colonne Vertébrale Simulant le Mal de Pott J radiol. et électrol. 6 21, 1922

[22] Sandstrom, C.. Calcifications of intervertebral discs and relation between various types of calcifications in soft tissue of the body, Acta radiol. 36:217, 1951

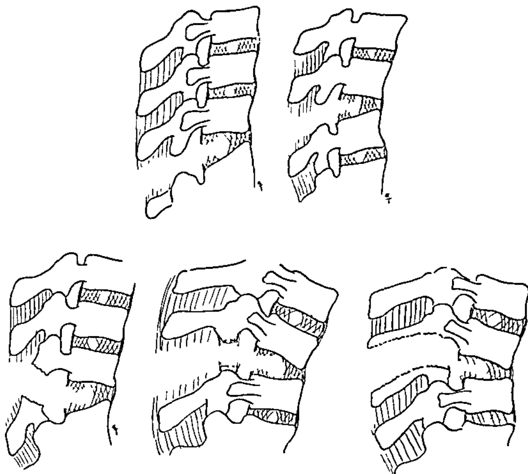


Fig. 490. Five types of injury to the posterior structures of the lower lumbar spine. (A) Springing of L 5-S1 space. (B) Springing of L 4-5 space. (C) Fracture through the pars interarticularis. (D) Fracture through the facet and springing of the L 4-5 space. (E) Longitudinal fracture through the spinous process. (Newman, P. H.: Sprung back, J. Bone & Joint Surg. 34B:30)

nulus fibrosus; (2) superiorly and inferiorly in the fibrocartilaginous plate, seen in roentgenograms as a thin transverse line; (3) centrally in the nucleus pulposus. The first two named are actually instances of metaplasia, i e., ossification in collagenous or cartilaginous tissue. The third named consists of deposit of amorphous calcium salts in degenerate tissue, not unlike the process of calcification in other situations where the tissue has undergone necrosis, e g., in the musculotendinous cuff of the shoulder. Similarly, the calcium deposit has the consistency of toothpaste and is capable of undergoing dissolution and resorption. Rarely, a solid accretion may form. Central calcifications are usually found in the adult spine, commonly the thoracic portion, rarely the lumbar, and are described as asymptomatic, existing perhaps for years before being discovered accidentally on a routine x-ray examination. However, the presence of the deposit implies that the disks in the area are degenerate and capable of extrusion posteriorly, thereby producing disk symptoms.[23] A disk as yet uncalcified may be responsible for symptoms, and one must depend on the clinically localizing signs and myelography rather than directly implicating the calcified disk. In drawing an analogy with the acute inflammatory appearance of the tendinous cuff overlying a symptomatic calcium deposit, it is not difficult to picture the same pathology in the annulus fibrosus. The latter structure contains the pain nerve endings. Thus may be produced local back pain followed by referred thoracic or abdominal pain as the annulus bulges posteriorly and then ruptures. When symptoms of local inflammation (back pain, local tenderness, muscle spasm, limited back motion) are present, treatment consists of recumbency, back support and x-ray irradiation (150 r every other day for 5 doses, using 180 Kv, filtration 1 A1 plus ½ mm Cu, 20 x 20 cm field)

Calcifications in intervertebral disks in children[24] are uncommon, but when they do occur they are accompanied by marked signs and symptoms. Typically, the condition is initiated acutely as an influenzal-like state with complaint of severe backache and a high fever. Erector spinae spasm and limitation of back motion is noted. Local tenderness is elicited over the affected disk. The sedimentation rate is increased, and a mild to moderate leukocytosis is present. Roentgenograms reveal the calcium deposit in the disk space corresponding to the site of tenderness. Several disks may be involved simultaneously. When a cervical disk is affected, stiff neck and headache are associated symptoms and suggest a diagnosis of meningitis. The acute symptoms generally subside in a few days to a few weeks on bed rest, and the calcium opacity disintegrates and disappears.

When many intervertebral disks are extensively calcified, the rare condition of ochronosis must be considered (q.v.).

SPONDYLOLISTHESIS

Spondylolisthesis is the forward slipping of a vertebra on a subjacent vertebra. It is most commonly associated with defects in the posterior bony arch, although vertebral displacement can occur as a result of malformation of the articular processes with lack of stability. When posterior arch defects are present but forward slipping has not occurred, the condition is called *spondylolysis* or *prespondylolisthesis*.

Etiology

The cause of the arch defects is unknown. Against a congenital origin is the fact that typical arch defects are almost unknown in the newborn. Nor can it be developmental, because double ossification centers on both sides of the neural arch in the fetus has not been found.[25] More plausible is a traumatic origin, possibly due to hyperflexion of the spine.[26]

Clinical Picture

Very insidiously, a deformity of the back gradually develops during childhood Rarely,

[23] Logue, V Thoracic intervertebral disc prolapse with spinal cord compression, J Neurol Neurosurg & Psych 15 227, 1952

[24] Walker, C S Calcification of intervertebral discs in children, J. Bone & Joint Surg 36B 601, 1954.

[25] Batts, M , Jr The etiology of spondylolisthesis, J Bone & Joint Surg 21.879, 1939.

[26] Hitchcock, H. H.· Spondylolisthesis. Observations on its development, progression, and genesis, J. Bone & Joint Surg. 22 1, 1940.

it appears acutely following a severe injury. The lumbar lordosis becomes exaggerated. The spinous process of the 5th lumbar vertebra forms a prominence above which is a depression. A transverse furrow in the soft tissues extends across the back at the level of the depression. For a variable period of time, usually many years, no symptoms are associated. Then often after the third decade, mild backaches appear spontaneously, following a severe trauma, or after a series of repeated lesser traumata such as an occupation requiring frequent bending and lifting. The discomfort is more pronounced with activity, the upright position, straightening up from a forward bent position, and hyperextension of the trunk. It is relieved by recumbency.

In many patients, no further disability develops. In others, the low back pain becomes progressively worse, is associated with occasional bouts of acute pain, muscle spasm and restricted back motion. Then gradually symptoms of nerve root irritation develop. Aching is perceived in the buttocks, the posterior thighs, the posterior and the lateral aspects of the legs, the heels and the feet. The discomfort is mild, moderate, or severe and may be unilateral or bilateral, in one region or the entire lower extremity. Some patients complain of coccygeal pain.

Hyperextension of the back is often associated with accentuation of radiating pain and sometimes causes pain about the coccyx. Forward bending may or may not be limited. Straight leg raising (Lasègue sign) is occasionally restricted on one or both sides and may aggravate the discomfort. This signifies adhesions between the 5th lumbar root and the pseudarthrosis at the defect in the pars interarticularis of the 5th lumbar vertebra It is not unusual to find a free range of back motion and a negative Lasègue sign Tenderness to pressure or palpation is observed over the prominent spinous process.

In some cases, signs of nerve root compression appear. In addition to severe sciatic pain and paresthesias, straight leg raising is restricted and is associated with increase of pain. Coughing, sneezing and straining aggravate the discomfort. Forward and backward bending are restricted. Most commonly implicated is the 5th lumbar nerve root by the defect in the 5th lumbar vertebra. Extension



Fig. 491. Spondylolisthesis. Note the attempt at formation of a supporting ledge at the anterior edge of the sacrum.

of the large toe is weak and sensation is diminished over the dorsum of the toe and the dorsomedial aspect of the foot. When the first sacral root is involved, the Achilles reflex is diminished, plantar flexion of the foot is weakened, and sensation is reduced over the lateral aspect of the lower third of the leg. Involvement of the 4th lumbar root by spondylolisthesis at the 4th lumbar vertebra results in a diminished patellar reflex and decreased sensation over the anteromedial aspect of the upper half of the leg.

When signs and symptoms point to nerve root compression, a ruptured disk must be considered in addition to a laminal defect. Most frequently (in 10 to 15 per cent of cases) a protrusion is observed at the 4th lumbar disk which compresses the 5th lumbar nerve root.

Roentgenologic Findings

A lateral spot view will reveal forward displacement of the vertebral body which carries forward the spine above it. Questionable slipping may be determined by *Ullman's sign:* a line drawn upward from the anterior surface



FIG. 492. Spondylolisthesis pathology. Dissected specimen to demonstrate the defects in the posterior structures. (A, *Top, left*) The upper arrows point to defects in the inferior articular processes. The lower arrow points to a defect between the sacral laminae where the spinous process should join the vertebra. (B, *Top, center*) The spinous process has been removed, revealing a spina bifida occulta defect in the sacrum. (C, *right*) The inferior segment of the right inferior articular process has been removed, revealing the sacral facet. Note the bony process of the remaining segment overhanging the nerve root as it enters the intervertebral canal. The root may be compromised as the 5th lumbar vertebra is displaced forward. *(Continued on facing page)*

of the sacrum normally lies at or in front of the antero-inferior angle of the 5th lumbar body. When the latter is intersected by the line, forward displacement has occurred. Routine oblique views reveal the typical bony defects in the pars interarticularis. The superior articular processes have a "puppy dog" appearance. The "neck of the puppy dog" corresponds to the pars interarticularis. In the *absence of displacement, defects in the pars* interarticularis indicate the presence of a spondylolysis. Displacement in the absence of laminal defects are associated with malformation of the inferior articular facets. The latter lie in a parasagittal plane, display degenerative changes and are displaced forward where they impinge against the upper border of the subjacent vertebra.

In the presence of restricted forward bending and straight leg-raising, the question of a disk protrusion requires myelography before surgical intervention.

PATHOLOGY[27, 28]

The characteristic defect is an interruption in continuity of the isthmus of the neural arch, usually of the fifth lumbar vertebra. The pseudarthrosis is narrow, obliquely disposed, and occupied by a fibrocartilaginous tissue. This tissue may extend into the spinal canal as a mass which compresses and frequently is adherent to neural structures. The pars interarticularis is elongated in proportion to the degree of forward displacement of the body of the 4th or the 5th lumbar ver-

[27] Adkins, E W O Spondylolisthesis, J. Bone & Joint Surg 37B 48, 1955.

[28] Gill, G. G, Manning, J G, and White, H. L.: Surgical treatment of spondylolisthesis without spine fusion, J. Bone & Joint Surg 37A:493, 1955.

Fig. 492 (*Continued*). (D, *Left*) The opposite inferior segment has been removed; here, too, the remaining segment overhangs and may compress the nerve root. (E, *Right*) A normal specimen after removal of the posterior arch shows the nerve roots lying free. (Specimens from Dr. Paul Milligan)

tebra. The intervening disk occasionally is ruptured or degenerate When bilateral laminal defects are present without forward slipping, the condition is known as *prespondylolisthesis* or *spondylolysis.*

In spondylolisthesis the intervertebral foramen, although distorted, is actually enlarged.

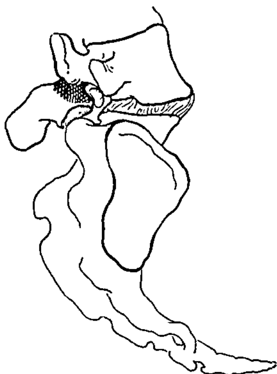


Fig. 493. Spondylolisthesis, diagrammatic. The defect occupied by fibrous or fibrocartilaginous tissue is represented by crosshatching. Note its intimate relationship to the 5th lumbar nerve root.

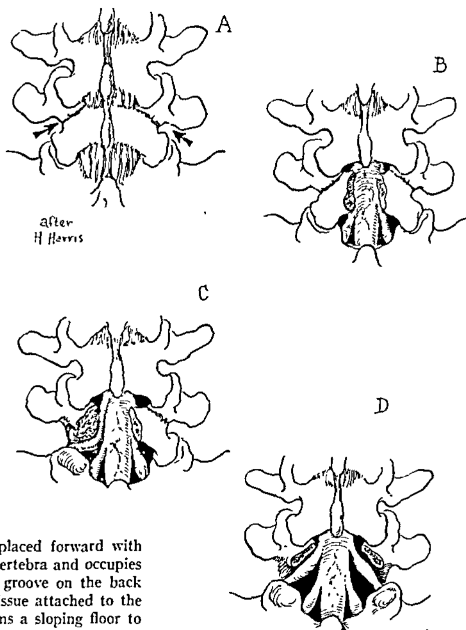


Fig. 494. Spondylolisthesis, diagrammatic representation of pathology. (A) Defect in the pars interarticularis of the 5th lumbar vertebra. (B) Relationship of 1st sacral root to the loose lamina. (C) Exposure of fibrous tissue in defect, after removal of loose lamina; as shown here, it may compress the 5th lumbar root. (D) Appearance after removal of loose laminae and fibrous tissue.

The nerve is always displaced forward with the body of the slipped vertebra and occupies its usual position in the groove on the back of this body. The disk tissue attached to the edge of the vertebra forms a sloping floor to this groove. The posterior wall of the upper part of the foramen is occupied by a fibrocartilaginous mass which is an inward extension of the tissue from the pseudarthrosis defect in the isthmus of the lamina. This mass often bulges into the foramen and compresses or is adherent to the root. If the lower margin of the slipped body forms an osteophytic outgrowth as a result of a degenerated disk, the groove occupied by the nerve root is narrowed. This narrowing in itself rarely constricts the root

The affected lamina is typically very loose and can be rocked through a wide range On extension of the spine it is rocked distally and pressed downward upon the fibrocartilaginous mass.

Spondylolisthesis can occur in the presence of an intact lamina.[29] It occurs most frequently at the L4-5 level in patients past middle life with severe osteoarthritis of the spine. The intervertebral joints are in a parasagittal plane instead of the normal oblique plane, and the upper facets subluxate forward. The articular cartilage on the facets displays extreme degrees of degeneration and erosion. Forward displacement of the lamina compresses the dura and causes an acute double bend on the dural sac between the lower edge of the lamina and the upper margin of the body of the next vertebra below. The forward displacement is limited by the inferior facets impinging upon the vertebral body below. In this situation the facet compresses the nerve root as it passes toward the pedicle and the

[29] McNab, I. Spondylolisthesis with an intact neural arch, J Bone & Joint Surg 32B:325, 1950

foramen at the next lower level. Usually the root becomes adherent to the displaced facet. The intervertebral foramen at the level of the slipped vertebra is usually unaffected. Compression of the theca and its contained roots can produce a typical cauda equina picture with sphincter weakness and saddle anesthesia.

When the dura is compressed between the lamina of the vertebra above and the posterior edge of the vertebra below, it is always indented and often densely adherent at both these levels.

The nerve root of the slipped vertebra always retains its normal relationship to the pedicle and the body of that vertebra. The nerve root of the next lower level usually emerges from the dura below the step but occasionally arises above it and is angulated over the sharp posterior edge.

The Cause of Symptoms

Instability and forward slipping of the vertebra is not the cause of severe symptoms There is no correlation between the degree of forward displacement and the degree of symptoms.[30, 31] Symptoms may often be relieved by removal of the lamina and the abnormal masses without spine fusion, thereby disproving the possibility that abnormal stress on fibrous or ligamentous structures may be the cause.

Because the lamina of the affected vertebra is very mobile, extension of the spine causes it to indent upon the fibrocartilaginous mass which in turn compresses the nerve root emerging at that level, usually the fifth lumbar. The lamina may rock downward and cause traction upon the first sacral roots, especially because the inferior margin of the loose lamina is often adherent to the dura. This causes coccygeal pain.

The actual cause of local symptoms is unknown.

Treatment

Cases with mild symptoms require rest, curtailment of activity, and a back support If disability is progressive and pain severe and persistent, surgical intervention is necessary. The procedures recommended are: (1) *decompression* by removal of the offending lamina and fibrocartilaginous mass. By this method, rehabilitation is rapid, and results are excellent.[32] (2) *Spine fusion.* (3) *Decompression and spine fusion.*

The pathologic process indicates that symptoms can be severe even in the absence of displacement. Restriction of forward bending and straight leg raising suggests incarceration of the nerve roots by adhesions, but a superimposed disk protrusion must also be considered. When coccygeal pain is a complaint and is accentuated by hyperextension, adhesions are to be anticipated between the inferior margin of the lamina and the dura.

Decompression and Spine Fusion. Regardless of which procedure is selected, the spinal exploration for causes of nerve root compression is mandatory. Conventional methods of spine fusion are often compromised by extensive removal of posterior arches. Therefore, when arthrodesis is contemplated, it can be attained by placing bone grafts from the sacrum along the lateral surfaces of the vertebral bodies and upon the transverse processes. A distracting "H" graft is placed between the spinous process of the sacrum and the one above the defect.[33]

Spinal fusion has been attempted by other methods. An interbody fusion is performed by inserting a bone graft through an abdominal approach. The surgical risk is considerable, and a pseudarthrosis often develops at the level of the disk. Interbody fusion may also be effected from the posterior approach by curetting out the disk and inserting small cancellous bone grafts. Extrusion of a bone fragment into the spinal canal is a definite hazard.

The following technic as described may be varied to suit the condition met with at operation.

Technic.[34] The patient is placed prone upon the operating table with supports to relieve abdominal pressure, thereby reducing the amount of bleeding. The middle portion of the affected lamina and spinous process is

[30] Friberg, S.: Studies on spondylolisthesis, Acta chir scandinav 82 (Supp 55), 1939.

[31] ———. Spondylolisthesis and trauma, Svenska lak.-tidning 36 379, 1939.

[32] Gill, G. G.: Personal communication.

[33] Bosworth, D M, Fielding, J W, Demarest, L, and Bonaquist, M Spondylolisthesis, J. Bone & Joint Surg. 37A:767, 1955

[34] Gill, G. G., Manning, J G, and White, H. L.. Surgical treatment of spondylolisthesis with spine fusion, J. Bone & Joint Surg 37A:493, 1955.

excised. The ligamentum flavum must be removed from the inferior aspect, especially to release the adherent dura. Similarly, the ligamentum flavum is removed superiorly. The lateral portion of the loose lamina is visualized in close relationship to the 5th lumbar nerve root. This portion is removed, first by disarticulating it from the sacrum, elevating it and dissecting it free from the defect in the pars interarticularis. The 5th lumbar root is dissected free and retracted medially. It is often adherent to the fibrocartilaginous mass and covered with large blood vessels. The fibrocartilaginous mass, when found, is removed until the nerve lies free. When the lower margin of the vertebral body displays a prominent ridge, the latter must be removed. Both 4th and 5th lumbar disks must be exposed and removed if found bulging or protruded. The superior articular processes do not require removal. Postoperatively, attention is directed to prevention of adherence of nerve roots. Rapid ambulation and straight-leg-raising exercises are necessary. An epidural injection of a solution containing hydrocortisone is helpful. The formation of adhesions is manifest clinically by a symptom complex: (1) attacks of catching pain in the back or the buttocks, (2) limitation of straight leg raising, (3) spasm on forward flexion with deviation toward the involved side, in order to avoid stretching of the affected nerve root, and (4) a list to the painful side when standing. This is an urgent indication for straight-leg-raising exercises and epidural injections.

If one wishes to add a spinal fusion, this is accomplished by laying bone grafts from the sacrum across the transverse processes and adjacent to the pedicles of the vertebrae. Then an "H" graft of cortical bone is inserted between the spinous process of the sacrum and that of the vertebra just above the affected vertebra.

Acute Spondylolisthesis. Forward displacement of a vertebra usually occurs slowly over a period of several years and most often at about puberty. Occasionally, the forward slipping may develop suddenly and to a severe degree. Reduction by manipulation may be possible. However, restoration of position does not preclude later development of symptoms, because of the mass of soft tissue at the isthmus defect.

Technic.[35] The patient is anesthetized. The head and the shoulders are supported on the edge of a table. The hips are flexed to the right angle, and then vertical traction is applied so that the lower limbs and the pelvis are pulled upward. The position obliterates the lumbar lordosis, the vertical traction pulls the sacrum forward, and body weight pulls the lumbar vertebrae backward. If necessary, a weight may be suspended across the abdomen in order to increase the backward pull. A double plaster spica is applied with the hips still flexed to the right angle. A few weeks later a spinal fusion is performed through a window cut in the cast.

Traumatic Spondylolisthesis. Severe injury to a normal spine can cause fracture of the pedicles or the articular processes, or dislocation at the facets. The vertebra dislocates forward, and compression of the cauda equina causes saddle anesthesia of the perineum and paralysis of the rectal and the bladder sphincters Reduction may be accomplished by the method described above. However, intact dislocated facets require open reduction. Otherwise, severe damage may be inflicted on the neural structures.

SPINA BIFIDA

Spina bifida is a defect in the posterior bony arch of the spine which may permit the meninges and their enclosed neural structures to herniate. Distortion of the spinal cord and the nerve roots results in neurologic deficits and deformities.

Embryology

The central nervous system develops from the ectoderm in the embryo. A groove forms longitudinally between two rising crests and progressively extends caudally. The folds converge dorsally and fuse to form the neural tube, the process beginning on about the 21st day. The initial site of fusion is at the caudal end of the hindbrain and is completed, at the cephalic pole of the neural axis, at about the 25th day and at the caudal end by the 29th day. Mesenchyme encases the neural tube at the same time. Therefore, this period of the 21st to the 29th days, when under the influ-

[35] Watson-Jones, Sir R. Fractures and Joint Injuries, ed 3, vol 1, p 331, Baltimore, Williams & Wilkins, 1944.

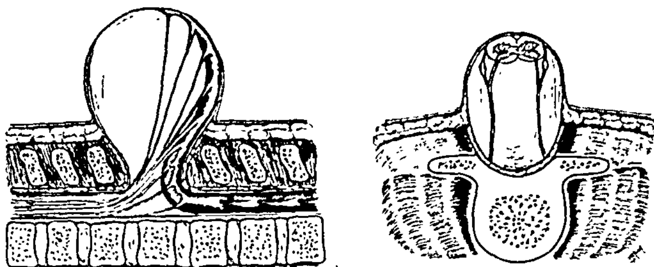


FIG. 495. Myelomeningocele. (*Left*) Sagittal section. (*Right*) Coronal section. (Campbell, J. B.: Congenital anomalies of the neural axis, Am. J. Surg. 75:231)

ence of various external factors, is prone to develop malformations of the spinal column and its contained neural tissues. It has been demonstrated that the normal process of invagination and fusion of the ectoderm is dependent upon proper mesenchymal encasement. Faulty mesenchymal formation results in failure of the tissue to fuse dorsally over the neuraxis and a bifid condition. In addition, other structures dependent upon the mesenchyme for their production, e.g., fat, blood vessels, fascia, ligaments, bones and some elements of the meninges, are abnormally formed and distributed about the bifid spine. The failure of the mesenchyme to act normally impairs the ability of the ectoderm to invaginate properly, and a myelomeningocele or an encephalocele is the result. (See section on "Congenital Anomalies.")

PATHOLOGY

The anomaly varies in degree from the very common *spina bifida occulta,* which consists of a small defect in the posterior arch, to a *myelomeningocele* with an extensive posterior bony defect. All types of combinations can occur, including skin manifestations, bony defect, herniation of meninges and neural structures, and formations of fibrous tissue, blood vessels, lipomatous masses and small bony deposits from the skin to the intrathecal cavity.

Spina bifida occulta consists of a small interruption in continuity of the posterior arch. It occurs frequently at the 5th lumbar vertebra or the upper end of the sacrum. The spinous process may be absent or is composed of two rudimentary processes from the laminae meeting but not fused in the mid-line. The intervening space is occupied by a fibrous membrane. Ordinarily, this is the extent of the anomaly and causes no symptoms. Occasionally, however, a fibrous band extends from the membrane outward, attaching to and retracting the skin. The fibrous process may also extend deeply, attaching to the dura and sometimes even penetrating it to engulf the roots of the cauda equina. In this case it often is fixed to the filum terminale, thereby restricting upward ascent of the spinal cord during growth. As a result, the hindbrain and the cerebellum are pulled into the foramen magnum (Arnold-Chiari malformation), and progressive spastic paralysis ensues. Constriction of the nerve roots at the level of the spina bifida may be mild in degree and effect slight subclinical muscle imbalance in the lower extremities which are manifest clinically as slowly developing foot deformity, notably equinocavovarus.

The following are the pathologic lesions of spina bifida:

1. *Bony Lesion.* This varies from a small mid-line defect occupied by a fibrous membrane to an extensive defect in which the laminae of one or more vertebrae are absent. The widest defects are invariably associated

with herniation of the meninges and the nerve structures.

2. *Meninges and Nerve Structures.* Meningeal protrusion through the defect constitutes a *meningocele.* When the spinal cord and the nerve roots are included in the protruding meningeal sac, the structure is known as a *myelomeningocele.* Some of the nerve roots are distorted and disorganized and end in the wall of the sac.

3. *Aberrant Mesenchymal Structures.* These include fibrous tissue, blood vessels, lipomatous masses and small bone formations. They are deposited anywhere from the subcutaneous area externally to the intrathecal space internally; they can occur independently or in any combination with each other. They are more likely to form profusely in association with a meningocele or a myelomeningocele and occasionally can develop as a mass in the subcutaneous tissue, bulging the skin outward and simulating a myelomeningocele. These aberrant tissue formations assume importance when the neural elements are impinged upon within the spinal canal. The lipomatous tissue can form a discrete mass pressing on nerve structures or may be diffusely distributed.

4. *Skin Lesions* The skin may be normal except for dimpling caused by a fibrous band. Other manifestations include pigmentation, angioma formation, hypertrichosis, and stretching and discoloration by the herniating sac.

Clinical Picture

The very common spina bifida occulta rarely, if ever, causes symptoms and is generally discovered accidentally on roentgenograms Occasionally, only a local area of pigmentation with hair formation is seen. Rarely, during the growth period of childhood, a spastic paralysis develops insidiously and signs of increased intracranial pressure appear. The feet develop equinocavovarus deformities Root pains in the upper extremities appear. This points to constriction of the cauda equina and restriction of the normal upward ascent of the spinal cord The hindbrain herniates through the foramen magnum. The cervical nerve roots assume a reversed course and are angulated. This is the picture of the Arnold-Chiari deformity, sometimes known as the filum terminale syndrome (q.v.). Milder degrees of the condition are manifest clinically as the gradual development of deformities of the feet and disturbances of gait.

The severe form of spina bifida with meningocele and myelomeningocele is apparent at birth. The newborn infant displays a large globular, dusky, tense, cystic tumor in the lumbar area. A profuse growth of hair overlies the mass. Both feet are clubbed. Hydrocephalus is often associated. Sphincter weakness is apparent.

Surgical Treatment

Spina Bifida Occulta.[36] Treatment is useless when major deformity and neurologic deficit exist at birth. Surgery benefits the patient born with minor deformity who shows increasing neurologic deficit as a result of interference with the dynamics of ascent of the spinal cord after birth.

By the 2nd fetal month, all coccygeal neural elements up to the first coccygeal nerve deteriorate and remain as the filum terminale. From the 3rd month on, the spinal column grows more rapidly than the cord, leading to ascent of the cord. At term, the tip of the conus medullaris arrives at the 3rd lumbar level. When a spina bifida occulta is present, a variable degree of distortion of the cord by adhesions and osseous and fatty masses will interfere with ascent and cause a neurologic deficit, the degree depending upon the severity of distortion. Therefore, neurologic disturbance may be absent at birth or may vary from a minor sensory, motor, or trophic change to a paraplegia or a quadriplegia, with sphincter incontinence. Commonest signs are underdevelopment of a foot or an extremity, valgus, varus, or cavus foot, or weakness in a major muscle group in a lower extremity. The commonest symptoms are disturbances of gait, station or posture Trophic disturbances and a sensory defect are seen. There is loss of control of bladder and bowel sphincters.

X-ray examination will reveal the extent of the defect. Pantopaque studies are necessary to define masses of bone or scar tissue which are adherent or impinge upon the cauda equina.

[36] Campbell, J. B. Congenital anomalies of neural axis, Am J. Surg 75:231, 1948.

Technic. Avertin rectally in the dose of 100 mg. per Kg. is the anesthetic of choice for children; open-drop ether, for the newborn. A mid-line incision extends between the spinous process above and that below the bifid area. The laminae of the normal superior and inferior vertebrae are exposed before the anomalous area is explored. Great care is exercised to avoid entering the dura and possibly the cord during the dissection. Every adhesive band should be suspected as a possible nerve root before it is cut. The findings vary tremendously. Anomalous osseous arrangements involving spinous processes, laminae, pedicles, facets and vertebral bodies are encountered in any combination. Commonly found are aberrant mid-line spicules of bone attached to the dorsal surface of a vertebral body impinging upon nerve roots, removal of which often results in reversal of clinical signs. Fatty malformations are subcutaneous, extradural and intradural. Decompression by laminectomy alone often ameliorates symptoms, even if the lipomatous infiltration of the cauda equina and the cord is diffuse. Fibrous tracts, which are probably sclerosed meningoceles or dural sinuses, lead down through the dura to the cord and are removed.

Meningocele and Meningomyelocele. Clinical differentiation between the two is almost impossible. If no neurologic disability exists, the lesion is probably a meningocele. The prevention of meningitis is paramount. The outlook is usually poorer when a myelomeningocele exists. The position, the size, the contour, the condition of the herniated meninges and availability of tissues for closure must be evaluated preoperatively Thin pedunculated meningoceles with a small stalk should be submitted to surgery as soon as possible. Rupture of the sac is an indication for immediate intervention. Unfortunately, the broad-based meningocele is more common. Inadequate tissue for closure and ulceration must await protective dressings of perforated cilkloid beneath which the epidermis grows and thickens Antibiotics are given to prevent meningitis. The age of choice for operation is between 12 and 18 months. Hydrocephalus or other outstanding deformities contraindicate operation. During the waiting period, contractures are overcome and prevented.

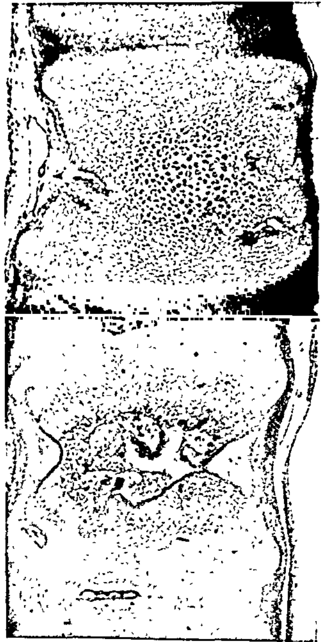


Fig 496. Vertebra of 14-week-old fetus. (*Top*) The body of the vertebra is composed chiefly of young connective tissue. Cartilage cells have begun to form in the center of the area. (*Bottom*) This is the first evidence of osteogenic tissue forming within the enlarging cartilaginous center. (Bick, E. M.: Osteohistology of the normal human vertebra, J. Mt. Sinai Hosp. 19:490)

Technic. An elliptical incision is made transversely to the neural axis and carried down to the deep fascia. The dura is opened at a point obviously free of neural contents, and the interior is inspected. Neural elements ending blindly in the wall of the sac are sacri-

ficed, and the intact structures are returned to the neural canal. The sac is resected, leaving only enough for water-tight closure. Dural closure is reinforced with fascia. If the spinal canal is too small for the replaced neural elements, a small reconstructed meningocele is created, and the superficial fascia and the skin are closed above it. Bony reconstruction is not attempted at this time. Postoperatively, the patient is placed in the prone position with the head down to reduce hydrodynamic strain on the suture line. Nursing care must be vigilant to observe spinal fluid leakage, decubitus formation and contamination of the wound.

OSTEOHISTOLOGY OF THE NORMAL VERTEBRA

For a proper understanding of diseases of the vertebrae, it is necessary to know the manner of histologic growth.[37]

In the early weeks of fetal life, the vertebrae composed of mesenchymal tissue have already assumed their form and are surrounded by the anterior and the posterior longitudinal ligaments. The intervertebral disks consisting of nucleus pulposus and annulus fibrosus are already in their proper positions. The mesenchymal tissue is transformed into cartilage, which becomes calcified preparatory to ossification.

At about the 15th week, blood vessels from the posterior surface penetrate the vertebral body at its center, carrying osteogenic tissue so that a focus of bone forms at the central portion of the body. At the earliest stages of bone formation, the trabeculae are chiefly vertically disposed.

[37] Bick, E M · The osteohistology of the normal human vertebra, J. Mt Sinai Hosp. 19 490, 1952.





Fig. 497. Vertebra of 25-week-old fetus. (*Top*) The ossific center has reached the periphery of the vertebral body. No appositional bone has appeared from the periosteum or perichondrium. Columnar cartilage makes its first appearance across the cephalic and the caudal surfaces of the ossific mass. These are the diaphyseal plates. (*Bottom*) Vertebra of 4-day-old female. At this age there is no difference between male and female vertebra. Observe the close apposition of the posterior longitudinal ligament to the ossific mass at the point of entry of the vessels. (Bick, E. M.: Osteohistology of the normal human vertebra, J. Mt. Sinai Hosp 19:490)

At about 25 weeks, the cartilage over the cephalic and the caudal surfaces of the ossific mass assume a columnar arrangement of cells adjacent to the bony centrum This is the earliest evidence of formation of epiphyseal plates by which the vertebral body grows in an axial direction. Because the vertebra does not actually possess epiphyses, the growth plate is more accurately termed a *diaphyseal plate*. The cartilage cells farthest away from the growth plate are flattened and characteristic of articular cartilage

At birth, the ossific mass is well formed and with the calcified cartilage on its cephalic and caudal aspects assumes the normal shape of the vertebral body. The calcified cartilage is gradually ossified until at about 3 months the ossific mass occupies almost all of the body and assumes the shape it will maintain throughout infancy and early childhood.

During these early years, the ossific mass occupies all of the vertebral body except a ledge about the margins of its cephalic and caudal surfaces. Instead, these depressions are occupied by cartilage which is continuous with the hyaline cartilage covering the superior and the inferior surfaces of the body. This ring of cartilage surrounds the rim except where it is deficient at the posterior end. Although it is thick, it forms only a slight elevation when the end surface of the body is viewed. The fibers of the anterior longitudinal ligament insert into the cartilaginous ring. Therefore, the latter is better termed a trac-

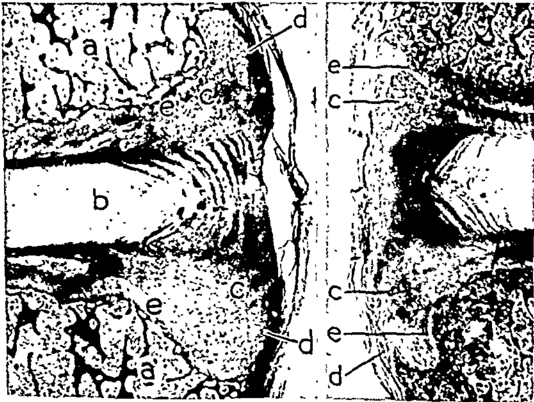
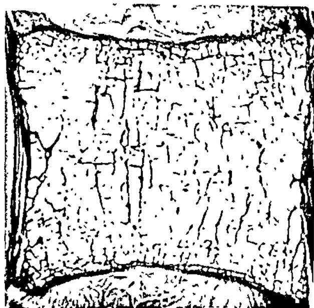


Fig. 498. (*Left*) Vertebra of 6-year-old female. This is the first appearance of the vertebral ring apophysis, as yet composed only of calcified cartilage. (a) Bone trabeculae of ossific mass; (b) nucleus pulposus; (c) calcified cartilage forming vertebral ring; (d) insertion of fibers of anterior longitudinal ligament; (e) diaphyseal plate (*Right*) Vertebra of 13-year-old female. This is the first evidence of ossification of the vertebral ring, seen at (c). Note that the ring lies outside the diaphyseal growth plate (e) and *is not in line* with the longitudinal axis of growth of the vertebral body. Fibers of the anterior longitudinal ligament are seen inserting into the area of the ring at (d). This insertion of the ligament into the accessory ossification center defines the ring as a traction apophysis. (Bick, E. M.: *Osteohistology* of the normal human vertebra, J. Mt. Sinai Hosp. 19:490)



FIG. 499. (*Top*) Vertebra of 18-year-old female The ,ring apophysis is now completely fused with the ossific mass. (*Bottom*) Vertebra of 80-year-old male. This section is stained by the van Giesen method in order to emphasize the trabecular matrix in bone substance The characteristic sparcity of trabecular substance in the aged is clearly demonstrated. (Bick, E. M.: Osteohistology of the normal human vertebra, J. Mt. Sinai Hosp 19·490)



tion apophysis, rather than the more common term of epiphysis.

The cartilage which lies central to the apophyseal ring is a thin layer of hyaline cartilage. As its cells approach the bony structure of the body, its cells assume a columnar arrangement preparatory to calcification, then degeneration and replacement by new bone. This is the true diaphyseal growth plate which contributes to the endochondral ossification by which the vertebra grows in length.

At 6 years of age, the cartilage in the apophyseal ring begins to calcify. This is the earliest age at which the notches at the anterior corners of the body as seen in roentgenograms display the triangular shadows of calcification. Within the next few years the calcification extends along the entire length of the ring and roentgenographically appears as a dense transverse line above and below the body.

Between the 13th and the 15th years in the female, and the 15th and the 17th years in the male, the apophyseal ring ossifies and fuses with the main ossific mass. Before this time, the unknown disease process which is characteristic of Scheuermann's disease interferes with proper calcification and ossification of the ring, which then appears as a vague interrupted line. In addition, the activity at the growth plate is interfered with, resulting in wedging. Similarly, curvature of the spine can be explained by asymmetrical interference with the growth plate. Conversely, the injurious forces are no longer effective after discontinuance of longitudinal growth Like other growth plates, the diaphyseal plate of the vertebra is subject to the so-called Hueter-Volkmann Law, which states in effect that pressure retards and traction accelerates the rate of growth.

At no time is the subchondral bone plate a continuous osseous surface. Instead, it is composed of flattened trabeculae interspersed with areas of marrow. It is a surface of spongy bone. Through these marrow openings cartilage under pressure of the nucleus pulposus may be driven through into the body to form the typical nuclear indentations of Schmorl's nodes.

The bony trabeculae of old age become thinned and lessened in number. The remaining trabeculae are disposed longitudinally.

Endochondral ossification at the growth plate continues but at a greatly reduced rate, supposedly because of lack of protein matrix upon which new bone can be laid down.

ADOLESCENT KYPHOSIS

(Scheuermann's Disease; Epiphysitis of the Spine; Juvenile Round Back; Osteochondritis Deformans Juvenilis Dorsi; Dorsum Rotundum)

A kyphotic deformity of the lower thoracic spine develops insidiously in individuals about the period of puberty. Often a history is obtained of indulging in unusually strenuous physical activity or participation in sports followed by the development of vague pain in the lower thoracic spine, radiating laterally toward both loins and subsiding with recumbency. Initial clinical and x-ray studies at this stage are unrevealing. Over the subsequent months, the pain diminishes, while at the same time the dorsal spine develops a gradual rounding or kyphotic deformity which is rigid and most pronounced at the lower thoracic spine. On attempting to bend forward, the lower thoracic rigidity is at once apparent, and the excessive forward bending of the spine above the area of involvement increases the kyphosis tremendously.

The almost exclusive *involvement of the lower thoracic spine* and its *rigid fixation* are the most characteristic findings. A lateral x-ray view now displays irregularity, mottling and *deficient ossification of the apophyseal ring*, especially anteriorly. The upper and the lower bony end plates, the most recently formed bone of the vertebral body, are blurred and irregular. *Small cuplike indentations* in these plates extend into the spongiosa within the body and become more apparent in time as the reactive ring of sclerotic bone forms about them. These are the typical Schmorl's nodes, formed by protrusions of disk material and consequently resulting in *narrowing of the intervertebral disk spaces*. The loss of disk cushioning results in increased flexion of the thoracic spine with increased pressure on the anterior portions of the vertebral bodies. Eventually, the *vertebral bodies become wedged* and narrowed anteriorly. The ring then completes its ossification and extends as a dense line, extending horizontally and parallel with the cephalad and the caudad surfaces of the body. When growth is complete after the age of 17, the cartilaginous growth plate between the ring and the epiphyseal bony end plate becomes ossified, and the ring fuses with the body. The deformity is now rigid and permanent. A peculiar characteristic seen in roentgenograms is the persistence of the transverse vascular grooves beyond the normal time of obliteration at age 6. The increase of the thoracic curve leads to development of compensatory increase of cervical and lumbar lordosis to maintain the center of weight-bearing. This eventually produces strain and degenerative changes later in life. This is seen best at the lumbosacral junction where the angulation becomes acute and forward pelvic inclination occurs.

Explanatory Anatomy

Longitudinal vertebral growth occurs from cartilaginous epiphyseal plates which cover the cephalad and the caudad surfaces of the vertebral body.[38] The process is similar to that in the long bones where columnar cartilage progresses into the osteoblastic area. This columnar formation disappears at 17 when subchondral bone plates form The proximal and the distal growth plates of cartilage are subject to the same deforming factors as in the long bones, namely, diseases, dyscrasias, metabolic or endocrine disturbances, asymmetrical pressures of posture or contraction, and x-ray irradiation. The epiphyseal ring is not an epiphysis, because it has no part in growth and therefore is misnamed.[39] This narrow cartilaginous mound (the "Randleiste" of Schmorl) lies on the rims of the cephalad and the caudad surfaces of the vertebral body. It more nearly resembles an apophysis of the traction type, its formation responding to attachments of the longitudinal and the intervertebral ligaments of the spine. Like other apophyses, when it approaches maturity, it fuses with the main body of bone. Its proper

[38] Bick, E M , Copel, J. W , and Spector, S.: Longitudinal growth of the human vertebra, J. Bone & Joint Surg 32A·803, 1950

[39] Bick, E M., and Copel, J. W.· The ring apophysis of the human vertebra, J. Bone & Joint Surg 33A 783, 1951.

ossification may be interfered with by the aforementioned factors. Ossification normally is apparent at age 6, and fusion with the body occurs at 17. When the apophysis is studied histologically, the ring is found to be deficient posteriorly and is thick anteriorly. It is the site at which branching fibers from the long intervertebral ligaments insert into the individual vertebra. These fibers exert traction at their insertion. The ring develops by a concentration of cells within the cartilaginous depression which surrounds the upper and the lower rims of the growing ossific mass of the vertebral body. Calcification of the ring occurs at 6, ossification begins at 13, and fusion with the body at 17. At 20 it cannot be identified and is covered over by the same articular cartilage plate which covers the end of the vertebra. At all times it develops outside the epiphyseal plate and contributes nothing to growth.

Theories of Pathogenesis[40]

Sequence of Events in the Disease. The actual exciting factor is unknown. The sequence of events suggests the following:

1. *Loss of disk substance* through defects in the cartilaginous end plate and into the body

2. *Excessive pressure* on the cephalad and the caudad surfaces of the body, mainly anteriorly

3. *Interference with ossification of the apophyseal ring* by pressure

4. *Interference with endochondral ossification* by pressure. Improper proliferation and maturation of the chondrocytes in the epiphyseal plate prevents normal bony replacement. This results in irregularity of the final bone end plates.

5. *Wedge-shaped vertebra develops* because the maximum pressure and retardation of longitudinal growth is anterior.

6. *Fixation of involved portion of spine* This is due to loss of disk substance and replacement by fibrous tissue

Explanation of the Disease. The following theories have been advanced to explain the disease:

[40] Schmid, P Zur Entstehung der Adoleszentenkyphose, Deutsche med. Wchnschr 74 798, 1949

A. *Loss of Disk Tissue.*[41, 42] Defects in the cartilage end plate, due to various causes, including nutritional and endocrine, permit the bulging disk to penetrate into the soft yielding spongiosa. The subsequent loss of cushioning against shocks of even normal weight-bearing and bending interferes with endochondral ossification. The axis of motion is posterior, and maximum pressure is transferred anteriorly where, therefore, restriction of growth is greatest and wedging occurs.

B. *Aseptic Necrosis.*[43] Excessive pressure on the vertebral body causes compression and gradual reduction in the caliber of the bony channels through which the vessels course. Reduction in circulatory flow results in aseptic necrosis, particularly at the ends of the body which succumb to pressure. This is more likely to occur when the body is weakened by persistence or enlargement of the vascular groove beyond the normal time of obliteration at 6. The proponent of this theory states that when vascular grooves are found in the adolescent who *is otherwise normal, wedging is* likely to develop, and prophylactic measures should be instituted.

C. *Endocrine.*[44, 45] Many patients display Fröhlich syndrome likenesses or exaggerated growth in length of the long bones, implying delayed closure of the epiphyseal lines. Under the influence of anterior pituitary growth hormone, growth is accelerated by increasing proliferation and maturation of chondrocytes in the epiphyseal plates. The plate becomes thicker and therefore more susceptible to compression forces. Injury to maturing cartilage interferes with proper ossification, resulting *in reduced rate of growth proportionate* to the degree of compression. This explains the re-

[41] Scheuermann, H Kyphosis Dorsalis Juvenilis, Z Orthop Chir 41 305, 1921

[42] Schmorl, G, and Junghanns, H. Die Gesunde und Kranke Wirbelsäule im Röntgenbild, Leipzig, Thieme, 1932

[43] Ferguson, A B Roentgen Diagnosis of Extremities and Spine, pp 350-360, New York, Hoeber, 1949

[44] Nathan, L, and Kuhns, J. G · Epiphysitis of the spine, J Bone & Joint Surg. 22 55, 1950

[45] Harris, W R The endocrine basis for slipping of the upper femoral epiphysis, J Bone & Joint Surg 32B 5, 1950.

duced height of the anterior as compared with the posterior portions of the vertebral body. The sex hormones, particularly estrogen, inhibit the effect of growth hormone

TREATMENT

Pressure on the anterior portion of the epiphyseal growth plate inhibits longitudinal growth at this site. Conversely, reducing pressure will permit the process to proceed normally. Assuming that sufficient growth time remains, the deformity of the vertebral body will be reduced or at least will not increase. If closure of the plate has occurred, further increase of deformity is impossible. These are the physiologic factors which form the basis for treatment. When the process is active, the patient is put at absolute bed rest in a position of hyperextension for a period of several months. Daily exercises are performed, designed to promote strengthening of the back extensor muscles. Then a body cast is applied to maintain the body in hyperextension, and the patient is allowed to be ambulatory. While in the cast, extensor exercises are continued. If adequate longitudinal growth of the patient has been completed, it may be possible to expedite closure of the epiphyseal lines by administration of sex hormones. Oral commercial preparations of combinations of androgen and estrogen are available, although parenterally administered preparations are more efficacious. A mild or moderate deformity may be accepted and causes practically no disability However, the more severe curves will cause obviously poor bodily posture and severe lumbar lordosis and lumbosacral angulation with its attendant symptoms. The deformity should be corrected by an extension jacket and the area fused surgically A spinal fusion will also eliminate symptoms of supervening degenerative arthritis at a later age.

Spastic Paraplegia in Adolescent Kyphosis.[46] When a previously asymptomatic patient with kyphosis dorsalis juvenilis develops a progressively increasing spastic paraplegia, the cause almost invariably is a spinal extradural cyst. The cyst typically arises in the lower midthoracic region and produces a severe spastic paraplegia with slight sensory changes and usually without pain or disturbances of sphincter function. Roentgenographic study reveals enlargement of the spinal canal and erosion of the pedicles. The diagnosis should be substantiated by myelography before surgical exploration.

[46] Cloward, R B, and Bucy, P C Spinal extradural cyst and kyphosis dorsalis juvenilis, Am J Roentgenol 38 691, 1937.

LOCALIZED OSTEOCHONDRITIS OF THE LUMBAR SPINE[47]

This condition is similar to osteochondritis of the thoracic spine. It occurs in the same age group, 10 to 16. However, the pathology is well localized to 1 or 2 vertebrae, whereas Scheuermann's disease is diffusely distributed over many vertebrae. Typically, the pathology consists of a punched-out defect at the upper or the lower anterior corner of a vertebral body. The disk space is narrowed. The body is slightly wedged. The opposing vertebral body presents a normal appearance. Clinically, *local pain and tenderness coming on* gradually over several weeks, referred pain usually at the hip, paravertebral muscle spasm and limitation of back motion are the main symptoms and findings. Differentiation from tuberculosis is important and is established by normal sedimentation test, normal differential leukocyte counts, negative Mantoux and absence of disease elsewhere. The condition rarely affects the cortex of the opposite boundary of the disk space. Pathologically, the microscope reveals an actual dissolution of bone or failure of formation of bone about the secondary ossification center (apophyseal ring)[48] The process is most pronounced at the anterior margin. Healing takes place when bone formation from the apophyseal ring proceeds normally once more, but a bony defect may persist to some degree. The latter is asymptomatic. Treatment consists of absolute bed rest for several weeks, followed by a back support. The prognosis is excellent for ultimate complete recovery over a period of 6 months to a year.

The condition is not comparable with a

[47] Lamb, D W · Localized osteochondritis of the lumbar spine, J. Bone & Joint Surg 36B 591, 1954

[48] deLorimier, A A The Arthropathies, p. 250, Chicago, Year Book Pub, 1943

Schmorl's node. The latter consists of a weakening of the end plate of the vertebral body and a breaking through of nuclear material into the body.

DEGENERATIVE JOINT DISEASE OF THE SPINE

Cartilage in the spine, as elsewhere, is subject to degeneration with advancing age. The loss of cartilage resiliency and disintegration varies in degree between individuals and seems to be favored by a hereditary tendency and by concomitant constitutional conditions including infection, menopause and metabolic errors. (For a full discussion on etiology, see section on "Affections of Joints.")

Pathology

Fibrocartilage of intervertebral disks degenerates and is replaced by fibrous tissue. Loss of cushioning effect between each pair of vertebrae leads to increasing pressures upon opposing surfaces of vertebral bodies which react defensively by increasing bone formation. Consequently, the bone hypertrophy produces increased density and irregularity, and outgrowth of bony spurs at the periphery. Occasionally, opposing osteophytes may completely bridge the interval and unite, thereby effecting an immovable juncture. Thinning of the disk anteriorly throws increasing stresses and pressures upon apophyseal joints posteriorly. The articular cartilage of the facets, already degenerate, is worn away. Facets which are subject to the greatest pressures and extremes of motion are most susceptible to deterioration. Therefore, the lumbar spine which supports the maximum body weight and is the site of much spine motion most often exhibits the greatest degree of degenerative changes. The cervical spine, because of its great range of motion, is the next favored site. When the direction of the facets is such that instability and excessive shearing occur, an increased degree of degeneration is invited The apophyseal joint interval is narrowed, and bony surfaces become sclerosed and irregular. The articular processes subluxate and overlap, the inferior facet displacing upward and encroaching upon the nerve root emerging at that level. At the anterior border of the intervertebral foramen lie the margins of the vertebral bodies from which spurs may compress the nerve root.

Atrophic (rheumatoid) arthritis may coexist with hypertrophic arthritis. The former has its onset earlier in life, and the loss of cartilage from articular surfaces quickly leads to hypertrophic changes. If, in the course of rheumatoid spondylitis, ligamentous calcification and obliteration of posterior articulations take place early, the spine becomes firmly fixed, and reactive hypertrophic changes do not ensue. When in an individual of advanced years one finds ligamentous calcification and fused apophyseal joints in addition to hypertrophic changes, one can assume that the course of rheumatoid arthritis has been slow and may yet be active.

Degenerative changes also affect the annular ligaments. Therefore, these individuals are particularly prone to disk protrusion. The consequent nerve root symptoms may be difficult to differentiate from root pain of foraminal encroachment.

Causes of Pain

Sprain of ligaments is common. The capsules are contracted about *rigid posterior* articulations and are easily disrupted by sudden forcible movement.

Irritative arthritis indicates an acute synovitis within the apophyseal joints in reaction to friction of irregular joint surfaces and the breaking off of minute fragments of cartilage.

Radiculitis is due to nerve root compression by an osteophyte, a subluxated facet, or swollen capsular tissues.

Disk rupture is common. The consequent nerve root symptoms may be difficult to differentiate from root pain of foraminal encroachment. The former usually has an acute onset and a tendency to improvement with recumbency. The latter appears gradually and becomes progressively worse.

Muscle spasm, when it is bilateral, occurs reflexly as a result of the aforementioned causes. Unilateral muscle strain may be caused by an unusually strong muscular contraction which is necessary for movement of the rigid spine.

FACTORS PREDISPOSING TO DEGENERATIVE ARTHRITIS OF SPINE

Local. Direct trauma to one region in the spine, or indirect trauma, such as a forcible flexion or extension movement which imposes

force upon some focal point, may eventuate in a localized degenerative arthritis.

1. Concussion. Small fragments of cartilage, not apparent roentgenologically, may be dislodged from articular surfaces and act as a foreign body within the synovial cavity of the apophyseal joint. Hemorrhage and edema of periarticular tissues encourage further degeneration of articular cartilage and subluxation of facets.

2. Fracture and Dislocation. Disruption of capsular structures and damage to articular surfaces is a natural accompaniment of these injuries.

3. Loss of Disk. Excessive strains and pressures thrown upon the posterior articulations encourage degeneration, capsular stretching and subluxation of facets. Experimentally, in animals, one can produce a localized degenerative arthritis of apophyseal joints by removal of the disk by an anterior abdominal approach.

4. Deformity of Vertebra. Regardless of the cause of deformity, whether congenital, traumatic, or infectious, the mechanical derangement will inevitably produce a localized degenerative arthritis.

General

1. Occupation. When requirements of an occupation include excessive carrying of loads and frequently repeated bending and lifting, particularly when carried to the point of fatigue, the joints of the lumbar spine are encouraged to deteriorate.

2. Mechanical or Postural. Proper balance distributed over the entire spine implies diffuse distribution of stresses and pressures. Muscle activity is held to a minimum, and fatigue does not easily set in. Improper posture demands excessive muscle contractions, fatigue appears quickly, and ligamentous structures bear the brunt of support. The facets subluxate, and their cartilaginous surfaces slowly disintegrate. Hypertrophic arthritis is inevitable in poor posture.

A deformity in one segment of the spine throws excessive strain upon another segment, with the result that the latter undergoes degenerative changes. For example, a dorsal round back or a swayback increases lumbar lordosis and imposes a heavy load upon the lumbosacral articulations which eventually reveal evidence of degenerative arthritis

Scoliosis, pelvic obliquity, and inequality of lower extremities similarly produce improper mechanics and osteoarthritis, particularly of the lower lumbar spine.

Obesity naturally encourages disintegration of articular cartilage in the lower articulations of the lumbar spine.

Antecedent Disease of the Spine. Any condition which directly attacks the integrity of the apophyseal joints, or indirectly by producing deformity of the vertebral bodies, will eventually cause degeneration of these joints. Vertebral epiphysitis which causes wedging, particularly of the lower thoracic vertebral bodies, invariably produces degenerative arthritis and stiffening of this segment of the spine. This rounding of the dorsum secondarily causes strain and degeneration of the lumbosacral joints.

Clinical Picture

Symptoms are both local and referred.

Local symptoms typical of degenerative arthritis include pain and stiffness at rest and are lessened by activity, heat and salicylates. The pain is intensified by unusual activity requiring excessive bending and lifting of heavy loads; it gradually improves with prolonged periods of rest. Symptoms are often worse during periods of cold, damp weather.

The discomfort may be intermittent or persistent, aching or sharp in character, localized or diffuse over the entire spine. When confined to one region of the spine, the areas of excessive movement (cervical spine) and of maximum weight-bearing (lumbosacral junction) are predisposed. Referred symptoms are typical of nerve root irritation and include pain, muscle spasm, paresthesias, etc. The dermatome affected by pain and objective sensory or reflex alteration defines the specific nerve root involvement. Girdle pains about the chest and the abdomen may simulate visceral disease.

Findings. The contour of the spine varies, usually depending upon an antecedent condition. Increased rounding and rigidity of the dorsal spine is associated with a greater degree of cervical and lumbar lordosis. The entire spine, particularly the lumbar portion, may be flattened, stiff and limited in motion in all directions. Forward bending is accomplished mainly by hip motion.

During a period of intense pain, reflex spasm may be noted in the erector spinae.

Tenderness to deep pressure or percussion over the spine may be localized to one site or distributed over a wide area.

Localized degenerative arthritis may be identified by: (1) *tenderness* over a focal point, and (2) by *local pain* intensified by a specific spinal movement. For example, backward bending will aggravate the pain of lumbosacral arthritis.

Intercurrent conditions which often aggravate symptoms include infection, metabolic disease, constipation, obesity and the menopause.

Roentgenologic Findings

The roentgenographic appearance is produced as a consequence of loss of articular cartilage from the facets posteriorly, and fibrocartilage from the disks anteriorly, bony hypertrophy of articular cortex being secondary.

1. **Apophyseal joints.** The joint interval is narrowed, the articular cortices are dense and irregular, margins are spurred, and the superior facets are often displaced downward and backward upon the inferior facets.

2. **Disk interval.** The spaces between vertebral bodies are narrowed (disk degeneration, disk rupture). The opposing cortices are dense and irregular and often contain radiolucent defects extending into the subcortical cancellous bone These are composed of disk material penetrating breaks in the end plates and are known as Schmorl's nodes. Osteophytic outgrowths (spurs) of varying size occur about the margins of the vertebral bodies and occasionally fuse with one another. Spurs over the posterior margins should be noted as a cause of nerve root irritation, particularly in the cervical region

3. **Deformities.** Wedging develops in the vertebral bodies, anteriorly in an exaggerated rounding of the thoracic spine, laterally in a scoliotic spine, and localized to 1 or 2 vertebrae following compression fracture Generally speaking, degenerative arthritis involving the entire spine tends to produce accentuation of cervical lordosis and thoracic kyphosis, and flattening of the lumbar curve

When calcification of the anterior longitudinal ligament fuses many vertebrae into a solid mass, its appearance appropriately suggests the name "bamboo spine." The condition most often follows rheumatoid arthritis, the degenerative changes being a secondary phenomenon.

Treatment

This consists of prevention, conservative treatment and surgical treatment.

Prevention. Direct trauma to the spine, whether or not fracture or dislocation has occurred, requires a period of *rest in recumbency* until hemorrhage and edema are obsorbed, ligaments regain their strength, and integrity of articular cartilage is preserved. *Fractures and displacements are reduced* so that proper spinal balance will not place articulations at a mechanical disadvantage *Change of occupation* or a reduction in working hours will prevent muscle fatigue and ligamentous strains. A program of carefully graduated *exercises* will increase tolerance for work. *Constitutional treatment* is aimed at correcting anemia, proper elimination, estrogenic and androgenic hormones, etc.

Deformity of the spine, whether caused by infection, neoplasm, fracture, scoliosis, epiphysitis, or congenital defect, produces mechanical imbalance that inevitably leads to degenerative arthritis. To maintain the erect position, an increased amount of muscle effort is required, fatigue sets in early, strain is thrown upon supportive ligaments which soon give way, and abnormal pressures and stresses are thrust upon articular cartilage. Cartilaginous tissue, which reasonably may be expected to survive the natural processes of degeneration until old age, disintegrates long before that time. *Maintenance of muscle power* is the key to retarding degeneration of the spine Therefore, it is not too surprising to observe patients with severe changes throughout the spine with a minimum of disability. On the other hand, a spine with a small degree of degeneration cannot remain asymptomatic in the face of activity which exceeds the limits of the musculature.

Conservative Treatment. Therapy which aims at relief of pain, increasing mobility of the spine, and strengthening the paraspinal musculature includes the following measures:

1. *Rest in Bed.* The position of greatest comfort is sought. As a general rule, slight flexion of the hips and the knees is best. A

firm mattress or a board under the mattress helps to reduce lumbar lordosis and relieves painful tension on articular ligaments.

2. *Heat.* Painful muscle spasm is reduced effectively by moderate warmth. Hot wet packs are laid along the spine and covered with waxed paper and an electric heating pad. Short-wave diathermy should be avoided, since it may cause necrosis of bony trabeculae which are already deficient in many of these patients.

3. *Massage* is best done following application of heat. It supposedly improves tone, circulation, and elasticity of muscles and effectively preserves integrity of the skin. Flooding the bed with a powder containing lanolin will reduce friction and prevent bedsores.

4. *Salicylates.* Sodium salicylate (gr. xv) is administered 3 times a day. Because it produces gastric irritation, the tablets should be enteric-coated and combined with an alkali. Salicylates are specific for relief of pain in osteoarthritis. Persistence of symptoms is cause for suspecting other pathology.

5. *Immobilization.* A corset, a brace, or a plaster cast provides rest to the spine in the ambulatory case. Supports are reserved only for the stage of acute pain. Their prolonged use is pernicious in causing muscle atrophy which in turn invites further degeneration of the spine.

6. *Traction.* Head halter traction is useful for cervical arthritis. Pelvic traction is applied in cases of lumbar arthritis. Both types of traction may be combined

7. *Exercises.* As soon as the acutely painful stage has subsided, general exercises are prescribed to strengthen the paraspinal, the abdominal and the gluteal muscles These must be carefully graduated and never performed to the point of fatigue.

8. *ACTH and Cortisone* and related compounds are indicated only when a rheumatoid arthritic component is present. Otherwise, their use is to be deplored as unnecessary and dangerous, particularly to patients of advanced age.

9. *Sex Hormones.* Occasional cases can be identified as developing during the menopause. At intervals, a repository type of androgen-estrogen mixture may be administered.

10. *Mineral Baths.* The benefits derived from a sojourn at a mineral spa include rest, heat and the laxative effects of drinking mineral waters.

Surgical Treatment. These measures include:

1. *Spinal Fusion.* When one localized site is the source of repeated disabling episodes, arthrodesis of the spine permanently relieves symptoms. The procedure is especially indicated for lumbosacral arthritis and the post-traumatic arthritis which follows fracture or dislocation, particularly in the cervical spine.

2. *Foraminotomy.* Compression of nerve roots is relieved by unroofing the foramina. The procedure is often combined with spinal fusion.

3. *Removal of Osteophytes.* Occasionally, a spur may develop at the posterior margin of a vertebral body where it impinges on neural structures. A favored site is the cervical spine where the osteophyte protrudes into the intervertebral foramen. This is a late sequel to a ruptured or degenerated cervical disk. Unroofing the foramen is usually sufficient for relief of symptoms.

4. *Removal of Ruptured Disk.* The advanced age of these patients is no contraindication to excising a protruding disk. Relief of symptoms is immediate, and rehabilitation is rapid. Otherwise, a downhill course is inevitable. Conservative therapy persisted in too long invites disaster. (See "Intervertebral Disk" in the section on "The Back.")

MARIE-STRÜMPELL ARTHRITIS[49, 50]

(Rheumatoid Spondylitis; Ankylosing Spondylitis; Bechterew Disease)

Marie-Strümpell arthritis is a definite disease entity, characterized by the occurrence in the adolescent of progressive inflammatory disease of the spine and the larger joints, particularly the shoulders, the hips and the knees, and leading to ankylosis and deformity. Definite clinical, pathologic and laboratory characteristics establish this condition as distinct from the spondylitis which is associated with rheumatoid arthritis, or that which is

[49] Kuhn, J. G.: Medical causes of backache, Am. Acad. Orthop. Surgeons, Lect., vol. 5, 1948

[50] Baker, L. D. Marie-Strümpell arthritis and the undiagnosed low back patient, Nebraska M. J. 33: 331, 1948.

superimposed upon a pre-existent osteoarthritis.

Etiology

The actual cause is unknown. Mechanical, infective, metabolic and endocrine factors have been blamed. The condition occurs predominantly in males. The late adolescent or the young adult is predisposed. These patients frequently have a slender build and poor posture.

Pathology

The microscopic appearance is indistinguishable from rheumatoid arthritis except for increased bone production adjacent to the joint. Grossly, the cartilage and the articular cortices are destroyed, and fibrous and bony bridging is frequent The para-articular tissues are degenerated and calcified, frequently ossified. The anterior longitudinal ligaments are especially affected by this process, the bony bridging between vertebral segments producing a rigid "bamboo spine." However, these anterior supportive structures are friable and easily ruptured by forceful extension of the spine. The aorta and the inferior vena cava which lie in close proximity to the spine anteriorly may be compressed and even ruptured by this sudden change of position. The apophyseal joints of the spine and the hip joints frequently are ankylosed by bone, the knees less frequently, and rarely the shoulders.

Clinical Picture

The condition starts insidiously in a young adult. Symptoms at first are vague and poorly localized. Aching and stiffness about both sacro-iliac joints occur as a morning backache which subsides with activity but returns after sitting in one position for long periods of time Sciatic radiation, unilateral or bilateral, is a common accompaniment. Rarely, the initial discomfort is about the lumbodorsal or the cervicodorsal area. The pain and the stiffness become progressively worse and spread slowly within 6 months to a year to the rest of the spine. Paraspinal muscle spasm is severe, the flexors predominating and pulling the entire spine into forward flexion so that the cervical and lumbar lordosis is obliterated, the thoracic rounding made worse, and a single kyphotic rounding of the entire back results. At the same time one or all of the hips, the knees and the shoulders become painfully swollen and distended with increased synovial fluid. The muscles acting about these joints undergo painful spasms which produce flexion adduction deformity at the hips, flexion of the knees, and adduction and internal rotation at the shoulders. At this stage the typical "stooped-over position" is evident. The patient gazes downward, the entire back is rounded, hips and knees are semiflexed, and the arms cannot be raised beyond a limited amount at the shoulders. Complaints include radicular pain in the upper extremities (cervical apophyseal joints involved), in the intercostal and the anterior abdominal areas (thoracic involvement), and in the lower extremities (lumbar involvement). Radicular pain in the groins is typical of sacro-iliac disease. The patient loses weight. Secondary anemia develops. On examination, in the early stages, when the sacro-iliac joints are involved, one can elicit tenderness over these joints either externally or on rectal examination. The Genslen sign is positive. Later, limitation of motion is detected at first in the lower spine, finally throughout the entire spine. Chest expansion is restricted because of disease of the costovertebral joints Endocarditis and iritis are frequently associated conditions. The initial acute inflammatory stage which occurs within the first 2 years is reversible by cortisone to a varying degree, depending upon the amount of destruction and ankylosis Otherwise, remissions are unusual. The disease is slowly progressive over 3 to 5 years. The sacro-iliac joints are fused, and the spine becomes ankylosed in a severe forward flexed position. The hips are fixed firmly in flexion and adduction. Usually, the knees and the shoulders have some residual motion. Pain subsides, radicular pain is absent, and a single rounded immobile spine curve results The vital capacity is greatly reduced, and respiration is diaphragmatic.

Laboratory Findings

Roentgenograms. Early, calcification and mottled trabeculation are found in the subchondral bone adjacent to the sacro-iliac joints, with haziness and apparent widening of the joint spaces. Changes are bilateral. Later, osteosclerosis advances, and joint fusion



Fig. 500. Marie-Strümpell type of spondylitis. The oblique view of the cervical spine shows ossification bridging the articulations posteriorly and disk spaces anteriorly, producing a straight immobile spine. Note that the foramina retain their size.

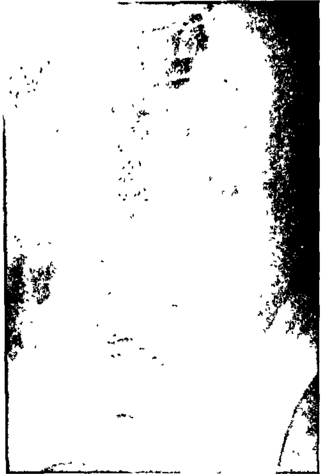


Fig. 501. Marie-Strümpell type of spondylitis. The oblique views of the lower lumbar spine reveal the earliest diagnostic changes: obliteration of the sacro-iliac joints, narrowing and obliteration of the posterior articulations, osteoporosis and (not shown here) straightening of the spine and ossification of the anterior longitudinal ligament.

occurs. The longitudinal spinal ligaments ossify, and the articular facets of the spine present haziness, destruction and bony fusion. The costovertebral joints may be fused. Hips display narrowing, osteoporosis, gradual loss of definition of articular cortices, and finally bony trabeculae bridging the joint

Erythrocyte Sedimentation Rate (ESR). This is elevated throughout the course of the disease and frequently the lifetime of the patient.

Urinary 17-ketosteroids are increased.

Secondary Anemia

Treatment[51]

Medical. This should be initiated and controlled in a hospital.

Cortisone is very effective in reducing inflammation and relieving pain. The deformity is overcome to a variable degree, depending upon the amount of fibrous and bony ankylosis. The method of administration is similar to that described for rheumatoid arthritis.

Roentgen Therapy. Relieves pain quickly, presumably by lessening muscle spasm. Relief generally lasts about 6 months. It is not curative. The following formula is recommended:

135 K V. 5 m.a.
12 in. distance
1 mm Al filter
75 r to each of 3 areas
12 treatments to each area
Treatment given every 2 or 3 days

In young women, treatment of the pelvic

[51] Baker, L D, Coonrad, R W, Reeves, R J, and Hoyt, W. A.: Marie Strumpell arthritis, J. Bone & Joint Surg. 32A 848, 1950



Fig. 502. Marie-Strümpell disease, involving both sacro-iliac joints. The joints are partially fused.

region must be avoided for fear of causing amenorrhea In spite of relief of pain, a decrease in range of motion and further ossification may occur. The bone condensations on either side of the sacro-iliac joints may clear up.

Recumbency flat in bed or hyperextended permits the spine to straighten.

Plaster Cast. A posterior plaster shell applied to the back while the patient lies in the prone position encourages quick reduction of inflammation and pain.

Deep-breathing exercises to preserve maximum vital capacity

Traction to cervical spine followed by a cervical collar as spasm lessens

Traction to lower extremities to overcome flexion deformities and distract joint surfaces. Theoretically, this may reduce the possibility of ankylosis

Hot, moist packs or a Hubbard tank aid in reduction of muscle spasm and permit strengthening of antagonist muscles.

Exercises, done after application of moist heat, are directed toward (1) back extensors, (2) hip abductors and extensors, (3) shoulder abductors and external rotators and (4) knee extensors

Well-molded plaster jacket is applied, maintaining slight lumbar lordosis and a straight thoracic spine. The Risser table is well suited for this purpose. Then ambulation is permitted.

Thorium X. Injections of thorium X have been given by Dr. Rudolph Wilde and Dr. C. Mau of Hamburg with complete relief of pain and what appeared to be permanent relief. Thirty injections were given at the rate of 2 per week until 6,000 units were used. The method is still experimental.

By adhering rigidly to the conservative program of treatment, it is possible to have the disease run its full course and yet have a minimum of ankylosis and deformity. However, sometimes in spite of treatment, the spinal and the hip deformities progress. The spine becomes a solid mass of bone from occiput to sacrum. The patient cannot see ahead, respiration is difficult, and disability is extreme. *A decision to correct and fuse the spine must be made early* before deformity is severe and endangers the aorta, the vena cava and the nerve trunks to the lower extremity.

Osteotomy of the Spine[52]

The procedure was introduced by Smith-Petersen in 1945.

Principles. A wedge of bone must be removed from the posterior elements of the spinal column, after which the spine is forcibly hyperextended It is done at the lumbar level (L2-3 or L3-4) because (1) the thoracic spine is fixed by rib attachments, (2) the lumbar

52 Adams, J. C · Technique, dangers, and safeguards in osteotomy of the spine, J. Bone & Joint Surg 34B 226, 1952.

canal is wider, and (3) it is done below the level of the cord. The fulcrum of the wedge is at the posterior rim of the selected intervertebral disk, i.e., the anterior margin of the corresponding intervertebral foramen. The angle of wedge is equal to the amount of correction.

Posture is lateral. This facilitates positioning of the deformed patient, allows full respiratory excursion, aids the anesthetist and eliminates the risk of injury to the ankylosed cervical spine.

Technic. Using a mid-line incision, spinous processes of 4 vertebrae, 2 above and 2 below the osteotomy site, are exposed. The site is determined before surgery by an x-ray marker. The ossified ligamentum flavum and parts of the adjacent laminae are excised. The dura may be abnormally adherent to the overlying bone and should be protected. After the dura is adequately exposed, a block of bone is removed laterally on each side from the lateral masses of bone comprising the superior and the inferior articular processes and the intervening joint which is usually ankylosed by bone. The wedge removed here is directed toward the front of the intervertebral foramen. It may be removed by a Kerrison rongeur (the underlying nerve being protected by blunt dissectors passed from within outward). An adequate amount must be removed to preclude the nerve root's being compressed when the spine is hyperextended. Then the spine is gradually hyperextended by pressure posteriorly on the manubrium sterni and the symphysis pubis and by anterior pressure about the osteotomy site This can be accomplished manually or by a simply constructed apparatus. The dura must be under constant surveillance during the correction. Straightening should be slow, to avoid sudden stretching of the abdominal structures. In the course of hyperextension, the anterior longitudinal ligament will be heard or felt to snap. When the wedge is closed, slivers of cancellous bone are packed into both gutters, and the wound is closed. A posterior plaster

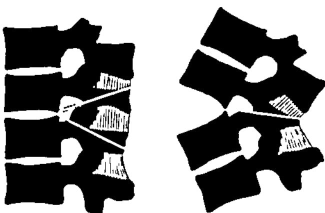


Fig. 503. (*Top*) Diagram showing the mechanics of spinal osteotomy. Note that the apex of the excised wedge is at the posterior margins of two adjacent vertebral bodies. (*Bottom, left*) Correct excision of posterior wedge. Its apex is at the anterior border of the intervertebral foramen. During hyperextension the fulcrum of movement remains in front of the cauda equina, which is consequently relaxed. The vertebral bodies remain in contact at their posterior margins. (*Bottom, right*) Incorrect excision of posterior wedge. Its apex is at the posterior border of the intervertebral foramen. During hyperextension the fulcrum of movement is behind the cauda equina, which is consequently strained. The vertebral bodies are forced widely apart. (Adams, J. C.: Technique and safeguards in osteotomy of the spine, J. Bone & Joint Surg. 34B:226)



shell is constructed at the conclusion of the operation. Later, an anterior shell is added. The patient is placed on a Stryker frame and is turned frequently. He is not allowed up before 8 to 10 weeks, and thereafter a body cast or brace is worn.

Dangers and Safeguards

1. ANESTHETIC DIFFICULTIES are overcome by the lateral position.

2 CAUDA EQUINA INJURY. *Stretching* is prevented by adequate removal of the posterior bone block so that the fulcrum will not fall posterior to the spinal canal. *Displacement or dislocation* during hyperextension or moving the patient is prevented by (1) excising bone so that bony apposition provides good stability, (2) correcting slowly under direct vision, (3) applying a full-length plaster cast

3. RUPTURED AORTA OR VENA CAVA. Theoretically, this is possible, especially if the aorta has degenerative changes. The aorta may be flattened anteroposteriorly. Extension should be gradual and through a small arc.

4. FRACTURE OF ANKYLOSED CERVICAL SPINE is prevented by utilizing the lateral posture.

5. ACUTE GASTRIC DILATATION AND PARALYTIC ILEUS. This is the greatest hazard in the early postoperative period. Prevention is achieved by immediate insertion of a Levin tube and administration of prostigmine. Treatment is by gastric suction, intravenous fluids, prostigmine and the prone position.

6. PRESSURE SORES develop easily. Turning frequently and flooding the bed with a powder containing lanolin will be effective.

Supplemental Surgery of the Large Joints

This follows the principles set down for rheumatoid arthritis As a rule, a joint partially or completely destroyed is not very adaptable for arthroplasty The para-articular structures and the muscles working about the joint are degenerate and replaced by scar tissue. Increase of motion to an acceptable degree is rarely obtained. Arthrodesis in a functional position provides stability and freedom from pain. The only indication for arthroplasty is bilateral hip involvement. If one hip is made movable, sitting, bending, climbing stairs, and other important functions are possible. In the knee, if flexion deformity and good motion are present, supracondylar osteotomy is best.

Flexion deformity of the hips will reduce the pelvic-femoral angle, throwing the rigid spine forward and causing an exaggeration of the stooped-over position. Before deciding on corrective osteotomy of the spine, it is necessary to establish to what degree flexion contractures of the hips contribute to the deformity. If the hip joints are not ankylosed, the Thomas test will measure the amount of persistent flexion in the hip. Correction is possible by release of soft tissue about the joint: i.e., capsule, rectus femoris, iliopsoas, adductors and iliotibial band. If fibrous ankylosis is present, arthrodesis in extension is necessary. In the presence of bony ankylosis, a subtrochanteric osteotomy corrects the flexion deformity. After the hips have been straightened, the degree of spinal deformity may be surprisingly small.

SPONDYLITIS OF RHEUMATOID ARTHRITIS

In typical rheumatoid arthritis, the peripheral small joints of the extremities are first *involved, and spread to the spine* occurs as a late manifestation. Either the sacro-iliac joints or the cervical spine may be the initial seat of the disease, spread to other parts of the spine is slower, occurring only during exacerbations, and crippling is not extensive. The differential diagnosis from Marie-Strümpell disease is as follows:

RHEUMATOID ARTHRITIS SPONDYLITIS

Both sexes affected
Primarily in peripheral joints. Spine is free
Poor response to roentgen therapy
Atrophy of subchondral bone
Acute onset, remissions and exacerbations
Serum agglutinates sheep erythrocytes.
Urinary 17-ketosteroids normal
Serum colloidal gold positive

MARIE-STRÜMPELL SPONDYLITIS

Males predominate.
Primarily in spine and large joints

Good response to roentgen therapy
Increased density of subchondral bone with mottling and loss of trabeculations
Insidious onset, slow progression
Agglutination tests negative
Increased excretion 17-ketosteroids
Serum colloidal gold negative

OSTEOPOROSIS OF THE SPINE

Osteoporosis of the spine implies a reduction in the number and the size of bony trabeculae which weakens the resistance of vertebrae to stresses and results in fractures, deformity and pain. The spine, the ribs and the pelvis exhibit the most prominent changes in osteoporosis.

Etiology

The main causes are the reduction of estrogenic influence at the menopause, senility, Cushing's syndrome, ACTH and cortisone treatment, immobilization and hyperthyroidism.

Pathogenesis

The processes of laying down of new protein matrix (anabolism) and resorption of old bone (catabolism) are constantly taking place throughout the lifetime. These processes are under the influence of endocrines, diet (protein availability) and activity (pressure stresses necessary for stimulation of new bone formation). The hormonal influence is particularly important. Sex hormones encourage the formation of new protein. A hormone elaborated by the adrenal cortex converts protein into glycogen and therefore is catabolic in nature. Any tumor of the adrenal cortex or a hyperplasia which is brought about by a pituitary basophilic adenoma or ACTH stimulation will effect protein destruction in this manner Senile osteoporosis seems to be a combination of several factors, including hormonal lack, inactivity, circulatory insufficiency and dietary and gastro-intestinal causes.

Pathology

Grossly, the cortex and the trabeculae of the vertebral body are thinned, and the marrow is fatty. The superior and the inferior bony end plates are concave from indentation by the expanded intervertebral disks, particularly in the lumbar area. In the thoracic portion of the spine, the bodies are compressed anteriorly, thereby accentuating the dorsal kyphotic curve. Microscopically, the transverse trabeculae are absent, while the longitudinal trabeculae are depleted in number and size. Osteoclasis is observed as normal in amount, but osteoblasts are noticeably sparse, and osteoid is absent.

Clinical Picture

Ill-défined aching pains about the back constitute the main complaint. A sudden, sharp pain may be provoked by a minimal trauma such as bending, lifting, or raising a window. The fragile vertebral body succumbs *to the slightest pressures, the fracture* varying from a crack through the cortex not demonstrable by roentgenograms to a severe compression of the entire body. Clinically, the affected portion of the spine is tender and, if compression is extreme, a kyphotic point is apparent. The surrounding muscles are in spasm. In the intervals between painful episodes, *the dorsal kyphosis is accentuated, and* the lumbar spine is flattened so that the patient appears to be "hunched over." Back motion is not greatly restricted. With repeated fractures, which are prone to occur about the dorsolumbar junction, the deformity becomes progressively worse.

Laboratory Findings

Blood Chemistry. The values for calcium, phosphorus and alkaline phosphatase are normal. An elevated glucose should direct one's attention to the pituitary and the adrenal as a possible cause of osteoporosis.

Other Studies. Determination of the urinary 17-ketosteroids (usually high), eosinophilic count (usually low), shift of the kidney shadow by I.V. pyelogram in the erect and the recumbent positions, and perirenal air insufflation may reveal the presence of an adrenal tumor. In postmenopausal osteoporosis, the urinary excretion of 17-ketosteroids is low, and calcium excretion exceeds that of intake B M.R. and radioisotope studies will reveal the presence of hyperthyroidism.

Roentgenologic Findings

In the thoracic spine the vertebrae are wedge-shaped, and the kyphotic curve is in-

creased. In the lumbar spine the expansile force of the disks is exerted upon the vertebral bodies at their centers so that their contour is biconcave. The disks are greatly thickened. The cortices are thinned and sharply outline the contour of the body, which appears homogeneous and washed out. An irregularity of the superior and the inferior cortical borders and anterior wedging suggest an old or recent fracture.

Treatment

Postmenopausal and senile osteoporosis generally respond to administration of sex hormones plus adequate diet. Activity is encouraged. Fractures need not be reduced nor do they require immobilization for fear of further depleting the bone Instead, a short period of bed rest is prescribed until the acute pain has subsided. Active back extensor exercises are instituted immediately and are continued indefinitely to promote new bone formation and as a defense against future injury.

During ACTH and cortisone therapy, the concomitant administration of androgenic sex hormone counteracts the catabolic effect of these substances.

Hyperthyroidism, basophilic adenoma and adrenal tumor require specific surgical treatment.

(See section on "Metabolic Diseases" dealing with "Osteoporosis.")

TUBERCULOUS SPONDYLITIS

Pathology

Any one or several vertebrae may be involved, the disease being most common in the lower thoracic and the lumbar spine. The infection starts in the cancellous area of the vertebral body where it may be mainly central, anterior, or epiphyseal in location. *Most commonly an exudative reaction with marked hyperemia produces severe generalized osteoporosis.* The body is softened and easily yields to compression forces. In the thoracic spine *the normal kyphotic curve increases the pressure* on the vertebrae anteriorly so that *anterior wedging is most severe in this region.* A pronounced angular kyphosis results if the body is crushed. In the lordotic cervical and lumbar regions, the center of gravity is thrown posteriorly, and the compression forces are less. Therefore, wedging is minimal.

The infection advances and destroys the epiphyseal cortex, the intervertebral disk and the adjacent vertebra. Or the infective exudate may spread anteriorly beneath the anterior longitudinal ligament to reach the neighboring vertebrae.

Infection of the posterior bony arch and the transverse processes is unusual. More commonly, granulation tissue develops posteriorly *and encircles and compresses the spinal cord* and the nerve roots. Pressure on nerve structures is more *likely* in the thoracic spine where the caliber of the spinal canal is small. Sequestra and bone fragments are rarely extruded into the canal, being *limited by* the strong posterior longitudinal ligament.

Anteriorly, the exudate forms and penetrates the periosteum accumulating beneath the anterior ligament. The exudate is composed of serum, leukocytes, caseous material, bone sand and tubercle bacilli. It penetrates the ligament and forms an abscess which migrates in various directions. It tends to track along lines of least resistance, i.e., along fascial planes, blood vessels and nerves.

In the cervical region, it collects behind the prevertebral fascia and spreads laterally to point at the posterior edge of the *sternocleidomastoid muscle. It may* protrude forward and bulge into the pharynx as a retropharyngeal abscess. The abscess may gravitate downward toward the mediastinum from which place it may enter the trachea, the esophagus, or the pleural cavity. When the origin is the thoracic spine, *the abscess* usually remains locally confined as a bulbous mass for a long time. Being thus confined, the back pressure is directed against the spinal cord, resulting in paraplegia. The exudate may spread *laterally to the extrapleural* space where its presence induces a nonspecific pleural effusion. If it penetrates the anterior longitudinal ligament, it occupies the mediastinum. Or it may gravitate downward through *the medial arcuate ligament* of the diaphragm to produce a lumbar abscess. Rarely, a thoracic *cold abscess may* track backward between the transverse processes and follow the intercostal nerve to erupt anywhere about the chest wall.



FIG. 504. Severe tuberculous kyphoscoliosis.

Tuberculous exudate originating from the lumbar spine usually enters the psoas sheath and gravitates downward to appear below the inguinal ligament on the medial aspect of the thigh. The exudate may spread laterally beneath the iliac fascia and emerge at the iliac crest at the anterior superior iliac spine If it follows the great vessels, it may erupt alongside the femoral vessels in the triangle of Scarpa, or in the gluteal region where it emerges with the gluteal vessels. Sometimes the exudate forms an abscess above the iliac crest posteriorly.

A single large caseative lesion of the body is rare. When it does occur, it remains isolated and calcifies centrally, appearing as a sequestrum. The structural strength of the body is not much weakened, and deformity does not occur. Involvement of the adjacent disk occurs slowly and late.

When multiple small foci of diminished density are present, this represents small areas of caseation and exudation. The mechanism of formation and spread of destruction is similar to the exudative form but is slower

The most common vertebral infection is the exudative type which constitutes a severe hypergic reaction, causes an extreme degree of osteoporosis, spread is rapid, abscess formation is frequent, and constitutional symptoms are pronounced.

CLINICAL PICTURE

Constitutional symptoms often antedate the local spinal involvement. These include weakness, anorexia, weight loss, night sweats and afternoon temperature. An exanthem or trauma frequently initiates the condition.

Symptoms. *Slight pain* and *stiffness* are the earliest complaints. The parents may notice a disinclination of the child to play. The pain is *localized* over the site of involvement or is *referred*, depending on the specific nerve root irritation. Cervical lesions cause pain in the occiput or the upper extremities; upper thoracic lesions produce intercostal neuralgia; lower thoracic infections cause discomfort about the abdomen and are frequently misdiagnosed as intra-abdominal in origin; lumbar tuberculosis causes pain referred to the lower extremities. As the infection progresses, *severe spasm* of the paraspinal muscles are the



FIG. 505. Pott's disease of the spine, showing destruction of the intervertebral disk and apposing bony structures, wedging and kyphosis.

source of severe pain Relaxation of the muscles during sleep permits painful motion which causes the child to awaken and cry (*night cries*). *Pressure effects* of the associated abscess causes additional symptoms: dysphagia, dyspnea, hoarseness (retropharyngeal abscess), dysphagia (mediastinal), and inability to extend the hip fully (psoas abscess) Stiffness, weakness and awkwardness of the lower extremities herald the onset of paraplegia.

Findings. The *attitude and the carriage* of the patient are very *protective* and careful Torticollis accompanies cervical disease. In upper thoracic involvement the shoulders are held high and back in military bearing. In lower thoracic and upper lumbar lesions the thorax and the head are thrown backward to produce an overerectness of the trunk. Lower lumbar lesions cause increase of lordosis and compensatory throwing forward of the chest to maintain balance.

During the early acute stage the *muscle spasm* may be so marked as to straighten out the involved segment of spine. As the spasm abates, the patient assumes the typical posture to counteract painful pressure on the anterior aspect of the vertebra and to maintain balance.

The back muscles rapidly undergo characteristic *atrophy*. Back *motion is restricted* in all directions, but particularly in forward flexion by muscle spasm.

Tenderness to pressure and percussion exists over the affected vertebrae. Tapping lightly over the spinous process frequently brings out the referred pain.

It is not uncommon to see a *boggy dusky thickening* of the soft tissues about the spine. This constitutes the paraspinal inflammation and may even herald the impending surfacing of a cold abscess.

Various areas should be investigated for *abscess formation*, including the posterior pharynx, the triangles of the neck, especially about the posterior border of the sternocleidomastoid, the iliac fossa, and the inner aspect of the upper thigh. Occasionally, the abscess may appear without antecedent complaints referable to the spine.

Paralysis of the lower extremities is spastic, and its appearance is denoted by hyperactive deep reflexes, positive Babinski, clonus, etc. Difficulty in emptying the bladder and the rectum signifies sphincter involvement. With further involvement of the corticospinal tracts, severe muscle spasm produces a flexion adduction position of both lower extremities. Rarely is the cord thoroughly destroyed. In such a situation the muscles become completely flaccid because of loss of reflex activity; generally, this is a result of some sudden accident, as a pathologic fracture dislocation. As the cord shock subsides, automatic sphincter actions and painful flexor spasms develop.

Kyphosis is an angular, sharp, local deformity caused by compression anteriorly of the body of one or more vertebrae. The dorsal protrusion may vary from a slight prominence of a spinous process to a marked projection. Compensatory lordosis occurs above and below the kyphosis. Lateral deviation of the spine is rare. A thoracic kyphosis causes the ribs and the sternum to be approximated to each other and displaced forward, resulting in a barrel-shaped chest. In a lumbar kyphosis the chest is displaced downward and forward, the lower ribs often coming in contact with the iliac crest.

Characteristics of Specific Spinal Involvement[53]

CERVICAL. Neck rigidity; pain over vertebra referred to occiput or arms, aggravated by pressure on top of the head; deformity—normal lordosis is reduced; child supports

[53] Mercer, W: Orthopedic Surgery, Baltimore, Williams & Wilkins, 1950

head in hands; retropharyngeal or cervical abscess; paralysis of arms before legs; occasionally, death due to dislocation.

LOW CERVICAL AND UPPER THORACIC. Marked rigidity with angular kyphosis; pain along brachial plexus; abscess—retropharyngeal, supraclavicular, mediastinal; cord symptoms less common, arms are affected first, Horner's syndrome due to pressure on sympathetic; neck rigidity, head and neck turn as one.

THORACIC AND THORACOLUMBAR. Marked angular kyphosis; girdle pain; pain referred to lower extremities, especially lateral aspect of thighs; iliac or psoas abscess.

LUMBOSACRAL. Deformity is slight; referred pain to lower extremities; psoas abscess; often flexion of hips.

ROENTGENOLOGIC FINDINGS

The acute exudative type being most common, the vertebral body becomes intensely decalcified throughout, the loss of density suggesting thorough destruction of the body. The margins, particularly those bordering on the disk, become indistinct. Gradually, the disk space narrows, signifying disk destruction. As the body recalcifies, anterior wedging compression is revealed. In the anteroposterior view, a dense shadow alongside the vertebra and extending distally is formed by the abscess. In the cervical region the abscess takes the form of a bird's nest. In the thoracic region it is bulbous and heart-shaped. Low dorsal and dorsolumbar abscesses are fusiform and occupy the site of the psoas shadow. Increased density and lessening in size of the abscess shadow denotes calcification and healing. Small or large caseous lesions of the vertebral body are less common and are revealed by localized osteolytic areas without reactive surrounding bone formation. Calcification of the central caseous substance imitates sequestrum formation. One or several adjacent or scattered vertebrae may be involved

TREATMENT OF TUBERCULOUS SPONDYLITIS

Measures are urgently needed to halt progress of destruction and deformity and particularly to prevent and possibly to overcome paraplegia. Immobilization, whether it be conservative by bed rest or surgical by spinal fusion, is most effective in improving the patient's general condition as judged by increased appetite and strength, return of temperature to normal, decrease in sedimentation rate, and lessening in size of the abscess. The most disastrous complication is paraplegia. It has never been known to develop after spinal fusion and often will subside after this procedure. Healing of the large bony focus is most effective by (1) spinal fusion, (2) drainage of the abscess, (3) excision of the focus and (4) combinations of antibiotics and chemotherapeutic agents. Eradication of this major focus lessens the possibility of dissemination and improves the patient's defensive powers so that other visceral foci may heal. Therefore, surgical attack is urgent. No attempt is made to overcome the deformity. Otherwise, the vertebrae may be distracted and create a defect in which the infection flourishes and is difficult to bridge by bony ankylosis.

Early Management. This consists of general measures (fresh air, high caloric and high protein diet, hygiene, vitamins), bed rest (hard mattress, Bradford frame, Stryker frame, posterior plaster shell), and combinations of streptomycin, para-aminosalicylic acid, and isoniazid or iproniazid. As the general condition improves, surgical treatment is instituted. Surgery is very urgent, even to the point of foregoing preliminary conservative treatment, when destruction is rapid, particularly in children, and when paraplegia is imminent or progressive.

Lymphocyte-Monocyte Ratio.[54] A high lymphocyte count and a low monocyte count is an indication of high resistance (a favorable allergic condition). Conversely, a low lymphocyte count and a high monocyte count is an index of low resistance (dangerous state of hypergy). The lymphocyte-monocyte ratio must be greater than 5 to have the most favorable conditions for surgery Complications are frequent, sometimes serious, when surgery is performed in a patient whose ratio is below 5. The test may be used instead of the sedimentation rate to determine progress.

Spinal Fusion. This is defined as surgically induced bridging of 2 or more vertebrae by a mass of bone. It should be performed early before major abscesses and paraplegia de-

[54] Campos, O P Bone and joint tuberculosis and its treatment, J. Bone & Joint Surg. 37A:937, 1955.

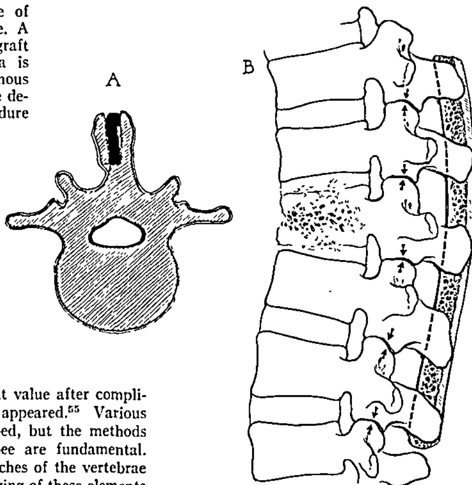


Fig. 506. Albee type of arthrodesis of the spine. A long corticocancellous graft removed from the tibia is wedged into split spinous processes. The facets are denuded. A Hibbs procedure may be added.

velop but is also of great value after complications have already appeared.[55] Various types have been described, but the methods of either Hibbs or Albee are fundamental. Because the posterior arches of the vertebrae are rarely involved, bridging of these elements by new bone is often successful. The kyphotic deformity should not be corrected. A draining sinus is no contraindication to fusion. In fusion of children longitudinal growth is not compromised. One or two vertebrae above and below the affected vertebra are included in the fusion. Before surgery a roentgenogram is taken with a metal marker over the suspected vertebral spinous process to determine the level of exposure.

Albee Procedure. The aim is to unite the spinous processes by a single cortical graft. The spinous processes are exposed through a mid-line incision, and the lateral muscle masses are elevated by subperiosteal dissection. The supraspinous and the interspinous ligaments are removed. The spinous processes

[55] Hallock, H, and Jones, J B: Tuberculosis of the spine An end result study of the effects of spine fusion operation, J Bone & Joint Surg 36A·219, 1954

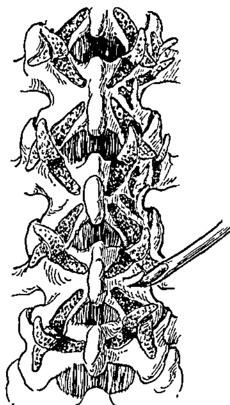


Fig. 507. Hibbs arthrodesis of the spine. Multiple slivers of bone are elevated from the sides of the spinous processes and the backs of the laminae and are crisscrossed over the interlaminar spaces.

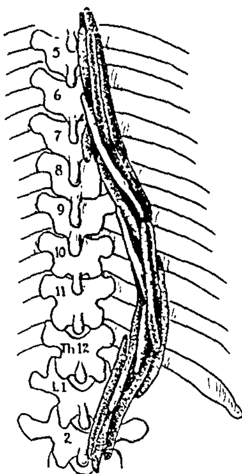


FIG. 508. (*Left*) Diagrammatic representation of circumduction fusion.



FIG. 509. (*Right*) Circumduction fusion of the spine. This is used when the neural arches are involved by tuberculosis or when an ill-advised laminectomy precludes the possibility of a mid-line fusion (Bosworth, D. M., and Levine, J.: J. Bone & Joint Surg. 31A:267-274)

are split longitudinally, and one half of each is fractured and displaced laterally. A bed of cancellous bone is thus exposed A full cortical graft is removed from the subcutaneous aspect of the tibia, placed in the gutter, and fixed by sewing the muscles over it.

HIBBS PROCEDURE. By this method only local bone is used. After exposure of the spinous processes and laminae, the supraspinous and the interspinous ligaments are removed The articular cartilage and cortex are removed from the lateral articulations by osteotomes and curettes. It is essential that a large bed of viable cancellous bone be exposed. Therefore, the spinous processes should be maintained. Multiple slivers of bone are removed from the sides of the spinous processes and the back of the laminae and are turned upward and downward to interdigitate with one another. Each sliver should remain attached at one end to preserve its viability A few small pieces of bone are packed into the defect left by removal of articular cartilage from the facets. The periosteum and the muscles are sutured snugly over the mass of bone.

COMBINED PROCEDURE. Fusion is more successful when bone chips are plentiful. The Hibbs and the Albee operations may be combined. In addition, cancellous bone chips may be obtained from the posterior superior iliac area, the spinous process beyond the area of fusion may be removed and split, or slivers may be removed from the tibia. These are packed into both gutters and the interspinous intervals. The more minute the grafts, the more rapid appears to be their incorporation into a growing fusion mass. Bone reinforcement is unnecessary in very young children.

Circumduction Fusion.[56] When the disease

[56] Bosworth, D M, and Levine, J: Tuberculosis of the spine, J Bone & Joint Surg. 31A:267, 1949

affects the posterior elements (neural arches), the infected field must be avoided. Also, when a previous laminectomy or a congenital absence of the posterior arch is present, fusion is difficult or impossible. For this contingency circumduction fusion is necessary. This consists of building a bony bridge laterally away from the pathology and extending between normal vertebrae above and below. The skin incision is lateral to the erector spinae, which is elevated. The ribs and the transverse processes are exposed and denuded of periosteum. Hemifusion with iliac chips and strips is done in stages, building from sound spinous and transverse processes above the tuberculous area outward onto the ribs, then down and inward to spinous processes below the diseased area.

Postoperative Care After Fusion. Prolonged recumbency for several months to a year may be necessary until consolidation is evident in roentgenograms. The Stryker frame permits ease of turning so that the patient spends equal periods in the prone and the recumbent positions. The wearing of a support, such as a brace, a corset or a plaster cast, seems to have no effect upon the progress or protection of fusion. A pseudarthrosis may develop and require refusion. This is more likely to happen over the apex of a kyphosis, particularly when an Albee tibial graft alone is used. The complication is least common in the lordotic cervical and lumbar regions where the lordosis provides a beneficial compressing force to the fusion.

The Abscess. Healing seldom occurs in the presence of a persistent paravertebral abscess.[57] The abscess is present in almost all cases, although it may not be visualized in roentgenograms. Fusion is an aid, but the dominant factor in healing is concerned with disappearance of the abscess material and its focus of origin.

In the pre-antibiotic era incision with drainage was contraindicated for fear of adding secondary infection with its consequent prolonged suppuration, amyloidosis and death. The boldest approach was aspiration by introduction of the needle through noninfected tissue in an effort to avert rupture. The advent of streptomycin has permitted open surgical drainage, following which tuberculous destruction of the vertebra ceases, and bone regeneration and spontaneous bony ankylosis

[57] Swett, P. P , Bennett, G E , and Street, D. M : Pott's disease, J. Bone & Joint Surg 22.878, 1940

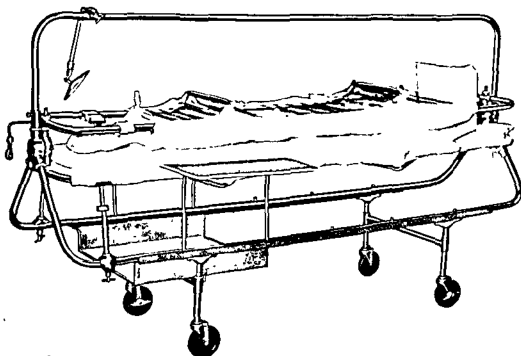


FIG. 510. Stryker frame. This facilitates turning the patient from the recumbent to the prone position, and vice versa.

frequently occur.[58] Streptomycin itself does not control the disease.[59]

An abscess which is small and confined, as in the thoracic region, causes back pressure, further destruction of the vertebrae, and encroachment on nerve structures. Given free exit, as when a large psoas abscess forms, destruction and nerve damage is lessened. Therefore, spinal caries with little or no evident abscess formation is an unfavorable combination. With progressive caries, the presence of paravertebral pus should be suspected, early exploration by lumbar transversectomy or thoracic costotransversectomy should be done, and the infective material cleaned out.[60] When the abscess is explored, the indolent scar tissue should be excised wherever possible, and a normal healthy bed of muscle, which is resistant to tuberculous infection, should be exposed. The infective bony focus and its fibrous wall should be removed and healthy surrounding bone exposed, the cavity being filled with bone chips. Cancellous bone is used because it is vascularized rapidly and does not sequestrate. Surgery is more urgent in the thoracic area where the danger of paraplegia is greatest and in children where destruction is rapid.

Relationship to Paraplegia.[61] The mechanism of production of most paraplegias is

[58] Deroy, M. S., and Fisher, H. Treatment of tuberculous bone disease by surgical drainage combined with streptomycin, J. Bone & Joint Surg. 34A 299, 1952.

[59] Stevenson, F. H. The chemotherapy of orthopedic tuberculosis, J. Bone & Joint Surg. 36B 5, 1954.

[60] Wilkinson, M. C. The treatment of tuberculosis of the spine by evacuation of the paravertebral abscess and curettage of the vertebral bodies, J. Bone & Joint Surg. 37B 382, 1955.

[61] Bosworth, D. M., Della Pietra, A., and Rahilly, G. Paraplegia resulting from tuberculosis of the spine, J. Bone & Joint Surg. 35A.735, 1953

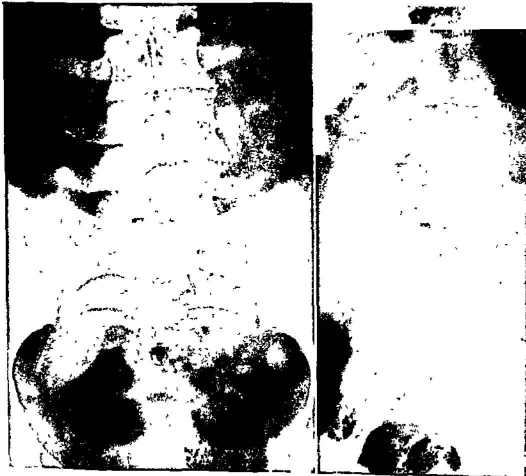


Fig. 511. Tuberculosis of the spine, 3 years after spinal fusion. Increased density and bony bridging of the interval between the bodies of the 4th and the 5th lumbar vertebrae attest to healing of the infected area.

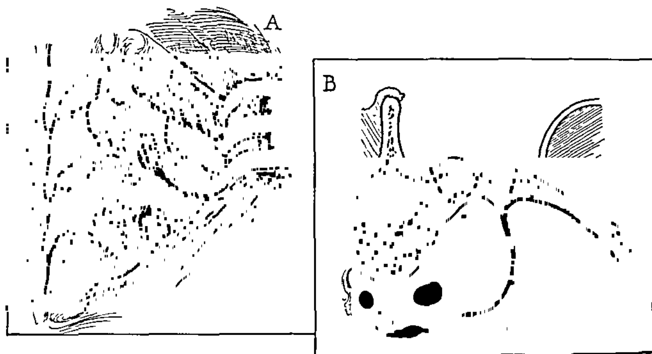


FIG 512. Costotransversectomy. (A) Exposure, showing extent of removal of rib and transverse process. (B) Cross section. The accumulation of abscess material extends laterally and compresses the lung.

unknown. The fact that, after drainage of an abscess, it frequently recedes even after existing for several years suggests that an edema due to toxins from neighboring exudate has a temporary inhibitory effect upon nerve cell function. Very rarely, direct pressure by a bony ridge, extruded sequestrum, chronic constrictive pachymeningitis and pathologic fracture dislocation are causative and are the only indications for laminectomy.

Prompt surgery is necessary to prevent a paresis from developing into a full-blown paraplegia. This consists of decompression of the abscess after spinal fusion or at the same time. Laminectomy is of no value in most instances. The outstanding early objective sign of paraplegia is sustained clonus Other signs of greatly increased pressure, such as a positive Babinski, weakness, and sensory changes, are found. Spinal taps must be avoided, because a meningitis may result.

Technic of Lateral Rhachotomy.[62] When definite spinal cord compression can be demonstrated, exposure of the anterior portion of the spinal canal is necessary. Capener described the technic as a means of decompressing the spinal cord without exposing the abscess to secondary infection. A curved incision is made to one side of the spine, extending several vertebrae above and below the lesion. It is deepened to the deep fascia, and the flap is retracted medially to the spinous processes. The trapezius muscle is incised at its origin and retracted laterally. At the level of the affected vertebra the erector spinae is divided transversely and retracted upward and downward. The rib is stripped subperiosteally from its angle to the head, and this section is removed For freedom of exploration an adjacent rib may be removed. The elevator costae muscles are excised. The intercostal vessels and nerve are located. The nerve is isolated and divided, the proximal portion being used as a guide for further dissection toward the theca. The pleura is exposed and depressed. The proximal end of the rib is rounded off to prevent its puncturing the pleura. The transverse process, the lamina and the pedicle are excised. Using the intercostal nerve for traction, the theca is elevated, and the posterolateral edge of the vertebral body may be visualized. Caseous material, sequestrum, degenerated disk and spurs can be removed. The wound is closed in layers.

[62] Capener, N · The evolution of lateral rhachotomy, J. Bone & Joint Surg. 36B 173, 1954

Technic of Costotransversectomy.[63, 64, 65] The exposure is the same as for lateral rhachotomy. Only the rib and the transverse process are removed. By blunt dissection to the anterolateral aspect of the body, the abscess and a ragged cavity in the bone may be encountered. The soft-tissue covering of the body is removed, and the interior is thoroughly scraped out until a freely exposed cavity with walls of cancellous bone is exposed. The cavity is packed with many small chips of cancellous iliac bone. All infective material is cleaned out. The wound is closed primarily, or a drain may be inserted for several days Often the wound heals quickly. Occasionally, a draining sinus forms but always closes in a few weeks. If the pleura has been punctured and a tension pneumothorax develops, insertion of a needle relieves the symptoms. The technic is easily adapted to removal of a psoas abscess and a lumbar focus.

OSTEOMYELITIS OF THE SPINE

Staphylococcus aureus is the usual offender, although any of the pyogenic organisms can cause acute osteomyelitis of the spine. When, as is frequently the case, the infection involves the vertebral body, the onset is insidious, and the course is protracted over several months. The bony trabeculae are destroyed, and the exudate extends upward or downward, destroying the intervertebral disk—posteriorly where it forms a peridural abscess and may cause a paraplegia, or anteriorly where it accumulates beneath the anterior longitudinal ligament. It may penetrate the ligament and involve the mediastinal structures in the thorax (empyema, suppurative pericarditis, mediastinal abscess) or in the lumbar region may extend downward as an iliopsoas abscess. It may extend along fascial planes and erupt anteriorly above the iliac crest or posteriorly alongside the spine or at Petit's triangle As the infection subsides within the vertebral body, an intense reactive bone formation occurs. This is noted within the first few weeks and is a differentiating clue against tuberculosis. The excessive bone proliferation forms huge exostoses and bridging between adjacent vertebrae.

Symptoms

Local symptoms include constant pain, particularly with forward bending. The overlying soft tissues are edematous, reddened and tender. Chest and abdominal pain indicates nerve root irritation or spread of the abscess. Meningeal signs, bladder weakness and paraplegia point to intraspinal abscess formation.

Less commonly, the infection is situated in the neural arch or its appendages. This is characterized by an acute onset with hyperpyrexia, excruciating tenderness, marked muscle spasm, and more intense edema, warmth and redness. Pain is severe.

Roentgenologic Findings[66]

Early, the body appears to be normal. Later, the disk space narrows, the trabeculae disappear, and the body collapses. Early, healing is evidenced by much reactive new bone with formation of huge exostoses and bridging. Rarefaction usually occurs at the center of the body or at the supero-anterior or infero-anterior angle. Demineralization is pronounced Neural arch infection is hard to detect except occasionally by oblique views

Treatment

Large doses of penicillin or chlortetracycline are given. If the causative organism can be obtained by biopsy aspiration or blood culture, it should be subjected to sensitivity tests and the appropriate antibiotic administered. An extradural abscess requires immediate laminectomy and drainage. When the body is destroyed, recumbency in a cast is mandatory. Upon healing, the vertebrae fuse spontaneously so that spinal fusion is rarely necessary.

Suppurative Psoas Abscess[67]

Purulent infection within the psoas sheath

[63] Menard, V · Etude Pratique sur le Mal de Pott, Paris, Masson, 1900

[64] Alexander, G L Neurologic complications of spinal tuberculosis, Proc Roy Soc Med 39·730, 1946

[65] Wilkinson, M C Curettage of tuberculous vertebral disease in treatment of spinal caries, Proc Roy Soc Med 43.114, 1950.

[66] Wear, J. E, Baylin, G J, and Martin, T. L.· Pyogenic osteomyelitis of the spine, Am J. Roentgenol 67 90, 1952.

[67] Zadek, I.. Acute non-tuberculous abscess, J. Bone & Joint Surg. 32A.433, 1950.

originates from acute or chronic osteomyelitis of the vertebral body in the lumbar region. The patient is acutely or moderately ill. Fever is high. Pain is referred to the pelvic fossa or the loin. Discomfort is experienced about the hip, the groin, the thigh or the lower back. Clinically, the hip is held in a flexed and externally rotated position Internal rotation is restricted and increases pain. The same is true of extension. While standing, the patient places the affected limb forward, the trunk is deviated toward the affected side, lumbar lordosis is increased, and the erector spinae on the opposite side is in spasm. Forward bending increases the pain and spasm. The pelvic fossa is tender Occasionally, a palpable mass is present. Roentgenograms may display an enlarged psoas shadow. Treatment consists of surgical drainage by a retroperitoneal exposure through a McBurney incision. Penrose drains are inserted and gradually removed. The appropriate antibiotic is given

Chronic Osteomyelitis of the Spine[68]

Chronic osteomyelitis like chronic bone infection elsewhere will repeatedly cause acute attacks of pain and fever, produce draining sinuses about the back and the abdominal wall, and in effect thoroughly invalid the patient. Amyloidosis is an ever-present danger. Treatment consists of exposing the vertebral body, thoroughly cleaning out all infective material and sequestrae, and packing the wound until filled by granulation tissue. Antibiotics are administered. Later, multiple small iliac cancellous bone chips are packed into the cavity, and a posterior spinal fusion is done. Recumbency over many months is necessary to prevent deformity.

NEUROARTHROPATHY OF THE SPINE[69]

(Vertebral Osteoarthropathy: Charcot's Disease of the Spine)

The condition of painless destruction, disorganization, excess ossification and hypermobility occurs in the spine comparable with neuroarthropathy elsewhere.

Etiology

The actual cause is unknown. Trauma to anesthetic joints is the presently accepted concept. Tabes dorsalis is present in most cases when the lumbar spine is involved Syringomyelia is associated when the lower cervical or cervicothoracic area is the site of pathology. Men in the 5th and the 6th decades are predisposed.

Pathology

The most mobile sections of the spine are affected, the lumbar and, less commonly, the cervical. One or more vertebral bodies exhibit (1) compression to one side with irregular sclerosis, (2) spur formations which are large, laterally disposed and may bridge 2 vertebral bodies, (3) fracture or melting away of half of the body, (4) large cystic cavities adjacent to the disk with sclerosis of the body, and (5) loose bone fragments anteriorly. The *intervertebral articulations are fragmented* Early, the disks are undisturbed; later, they are destroyed. Dislocation lateralward and backward is frequent.

Clinical Picture

Trauma often initiates the disease, following which the condition appears to develop rapidly over a short period of time. Pain is minimal or absent, but weakness and instability *are prominent complaints.* On examination, a kyphosis or kyphoscoliosis of the lumbar or the thoracolumbar spine is apparent. New bone formations or irregularities may be palpable in the tissues about the spine. No tenderness exists. Although the affected segments are rigid, the remainder of the spine exhibits movements which are excessively free so that the individual is capable of placing both palms on the floor without bending his knees. A sharp thud is often palpable and audible on flexion-extension movement. Other large joints may be afflicted.

Roentgenologic Findings

Gross destruction, disorganization and massive new bone formation are found *A vertebra* may be subluxated laterally or posteriorly. The osteophytes are large and beaked. Disk

[68] Wiltberger, B. R: Resection of vertebral bodies and bone grafting for chronic osteomyelitis of the spine, J. Bone & Joint Surg 34A 215, 1952

[69] Thomas, D. F Vertebral osteoarthropathy J. Bone & Joint Surg 34B 248, 1952

spaces are narrowed. The bone ends beneath the articular cortex are sclerosed. Spinous processes are thickened. Vertebral fractures occur.

Treatment

Protection by a brace or a corset is wholly inadequate. A spinal fusion extended beyond the limits of the involved area provides stability and prevents further destruction.

FIBROSITIS
(Myofascitis; Lumbago)

Localized painful, tender, indurated nodular deposits may form in fascial sheaths enveloping muscle and cause diffuse, ill-defined back pain. The condition usually affects individuals of middle age. Pain is often initiated and aggravated by overuse of the affected muscle, exposure to cold and drafts, and focal sepsis. It is accentuated by rest when it is associated with stiffness. Heat and exercise temporarily relieve symptoms. On palpation, small sensitive nodules are found, or a crepitant sensation accompanies active contraction of the muscle. Infiltration with a local anesthetic relieves the pain, whereas injection with hypertonic saline intensifies the local and referred pain. The cause and the actual pathology are unknown. The condition must not be confused with painful infiltrations of rheumatic fever.

The most common sites are about the posterior iliac crest, either above or below, the supraspinatus area of the scapula, the lumbar portion of the erector spinae and the trapezius Nodules about the iliac crest often cause referred pain to the sciatic area Forward bending places tension upon the affected muscle, reproducing the discomfort Supraspinatus fasciitis causes pain referred to the shoulder. Abduction of the arm intensifies the symptoms Involvement of the upper half of the trapezius is usual and often causes pain referred to the cervical area. Limitation of forward bending of the neck is due to stretching the painful muscle. Any muscle may be similarly affected

Some writers include as fasciitis the painful syndromes found at the musculotendinous attachments. Examples are· the lateral epicondyle of the elbow (common extensor tendon), superior nuchal line of the occiput (posterior cervical muscles, a common cause of headache), and the twelfth rib (quadratus lumborum). However, these conditions are better interpreted as partial avulsion or chronic strain of the periosteal attachment.

Painful lipomata about the lower back need not be confused, although treatment is similar. These fibrofatty tumors are softer, lobulated and, instead of being fixed to a fascial sheath, slip about beneath the palpating finger.

Treatment

The nodule is destroyed by multiple punctures with a large-bore needle under local anesthesia. The muscle is exercised constantly to prevent painful adhesions. Soreness resulting from the trauma lasts only a day and disappears with the original symptoms. Deep kneading massage after heat applications has been recommended as a means of breaking up the nodules, but this is time-consuming and often ineffectual. Foci of infection must be eradicated.

Vitamin E orally in large dosages may be given to prevent recurrence. Steinberg has successfully treated fibrositis by administering the mixed tocopherols over several months.[70] The recommended dosage is 300 mg. of mixed tocopherols daily for the first week and 150 mg. daily for the next 2 weeks. A maintenance dose of 100 mg. daily is continued thereafter. This suggests its possible benefit in other conditions with fibrous proliferation, e g., muscular dystrophy and Dupuytren's contracture.

VERTEBRA PLANA
(Calvé's Disease of the Spine)

In 1924 Calvé first described a condition of the vertebral body which he called "a localized affection of the spine suggesting osteochondritis of the vertebral body with clinical aspects of Pott's disease." The condition is characterized by rapid collapse and flattening of a single vertebral body which appears as a dense wafer in roentgenograms. Over several years the body regains its original normal density and a variable portion of its height. The roentgenographic appearance suggests an

[70] Steinberg, C L: The tocopherols (vitamin E) in the treatment of primary fibrositis, J. Bone & Joint Surg 24 411, 1942

aseptic necrosis with collapse due to weight-bearing and regeneration by creeping substitution. However, recent biopsy study of similar lesions indicates that the cause may be an eosinophilic granuloma.[71] The increased density may be due to compression of the undestroyed remaining trabeculae. The body regains some of its original height by continuing growth after the lesion has spontaneously healed.

Clinical Picture

The onset is insidious, with the appearance of slight-to-moderate pain, fatigue, night cries, muscle spasm, tenderness well localized over the affected spinous process, and the gradual development of a kyphosis or scoliosis. Children are affected. The symptoms subside over a period of about 2 months, but the deformity remains.

Roentgenologic Findings

One vertebra is usually involved. At first, the body is slightly wedge-shaped and as it becomes progressively more compressed it assumes greater density. Finally, a flattened waferlike segment is seen with an associated deformity of the spine. The adjoining disks are normal. Over a number of years, the body height is moderately restored, and its internal architecture becomes normal.

Treatment

Recumbency may retard the degree of compression. Based on the assumption that an eosinophilic granuloma is present, x-ray therapy may effect a cure. To prevent deformity, it may be necessary to perform a spinal fusion. This does not interfere with longitudinal growth of the spine. It is not advisable to attempt to remove the lesion. The prognosis is favorable.

Differential Diagnosis

Confusion with tuberculosis is possible in the early stages. The sedimentation rate is normal. The disks are unaffected. A tuberculin test should be done. When in doubt, biopsy is indicated, either by needling or by direct surgical approach.

[71] Compere, E L, and Coventry, M B.: Vertebra plana due to eosinophilic granuloma, J Bone & Joint Surg. 36A:969, 1954.

ASPIRATION BIOPSY OF LESIONS OF VERTEBRAL BODIES[72, 73]

Most lesions occur in the vertebral bodies anteriorly where they are inaccessible for biopsy. However, a sufficient amount of tissue can be removed through a large-bore needle inserted from the posterior or the lateral aspect. The procedure is recommended only for the lower 4 cervical, the lower 4 thoracic and all of the lumbar vertebrae. The upper 3 cervical can be reached by the pharyngeal route. The upper thoracic vertebral bodies are closely covered by the parietal pleura, descending aorta and vena cava; this area is better exposed directly rather than explored blindly. In the lumbar region, the large vessels lie anteriorly where they are unlikely to be damaged.

Technic for Lower Thoracic and Lumbar Regions

The patient is placed in a prone position on the x-ray table. A calibrated rod is laid over the spinous process perpendicular to the skin. A lateral roentgenogram is taken, and the distance from the skin to the anterior border of the vertebral body is measured, using the shadow of the measuring rod to compensate for distortion. A simple trigonometric equation determines the distance that the needle must be inserted from the mid-line in order to strike the vertebral body. Also, the depth of insertion is calculated. The calibrated rod is removed, and the skin is prepared. A wheal of local anesthetic is made over the tip of the spinous process and another lateral to the spinous process at a distance previously calculated. At the latter point, a small incision is made to permit insertion of the needle without clogging the tip with skin. The Valls guide is placed in position. This consists of a centimeter rule at one end of which is a guide channel fixed at 55°. The sliding centering pin is fixed at the required distance and is placed over the spinous process. Then the guide channel should be in accurate approximation to the lateral incision. A

[72] Ray, R. D.: Needle biopsy of lumbar vertebral bodies, J. Bone & Joint Surg 35A:760, 1953

[73] Valls, J, Ottolenghi, C. E, and Schajowicz, F: Aspiration biopsy in diagnosis of lesions of vertebral bodies, J A.M.A 136:376, 1948.

14-gauge needle and its sheath are inserted through the guide channel and pushed inward until they come in contact with the bone, and roentgenograms are taken to confirm the position. The needle is withdrawn, and the trephine on the end of the sheath is used to remove the biopsy. As the trephine is removed, constant suction is applied by a syringe attached to the sheath to ensure against loss of the tissue. Then the tissue is expressed from the trephine by an obturator and placed in saline.

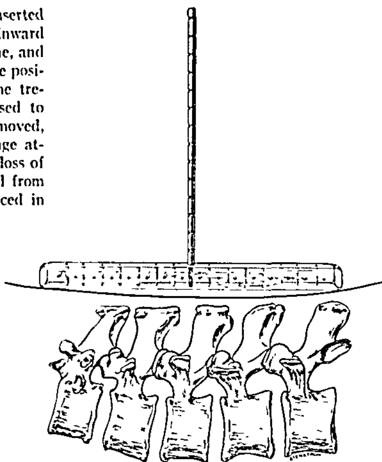


Fig. 513. (*Top*) Needle biopsy of the vertebral body. The metal ruler and rod are calibrated at half-centimeter intervals The affected body is located, and the distance from the skin to the anterior border of the body is determined. (*Bottom*) Using a trigonometric equation, the distance the needle must be inserted from the midline (AC) is calculated. The distance from the point of insertion to the anterior portion of the body (CB) is also calculated The Valls guide, sheath and needle are shown in place. (Ray, R. D.: Needle biopsy of lumbar vertebral bodies, J. Bone & Joint Surg. 35A: 760)

A C 55° B

$AC = \frac{AB}{\tan 55^\circ}$ $CB = \frac{AB}{\sin 55^\circ}$

$AC = \frac{AB}{1.43}$ $CB = \frac{AB}{.82}$

⑪ ⑩ ⑨ ⑦ ⑤ ④ ③ ②

STENSTROM

Technic for Lower Cervical Region

A vertical line is dropped from the tip of the mastoid process. The needle puncture is made laterally behind this line, thereby avoiding the large vessels and nerves which lie beneath the sternocleidomastoid. The needle is pushed medially until it comes in contact with bone, and confirmatory roentgenograms are taken. If the vertebral artery is penetrated, the needle is withdrawn slightly and directed a little anteriorly. During insertion of the needle, constant suction by syringe is exerted to detect penetration of a vessel. It is impossible to enter the spinal canal because the foramina have an oblique direction On bony contact, the trephine is pushed inward after the stylet needle is withdrawn. While removing the trephine, syringe suction will prevent loss of the tissue.

Upper Thoracic Region

Lesions of the upper thoracic bodies should be exposed surgically by removal of a transverse process and approaching the body under direct vision. The procedure is more formidable but has the advantages of exposing and avoiding important structures, draining and curetting out inflammatory material and affording an opportunity for fusing the spine immediately.[74]

POSTURE: NORMAL AND ABNORMAL[75-78]

Definition

Posture is the positional relationship of the regions of the body to each other. The total configuration of the body is the sum total of the contour of the spinal column and the contour of the lower extremities. Posture may be divided into standing, sitting and recumbent positions. The following description applies only to bodily contour in the orthograde position.

"Normal" posture is generally accepted as follows: moderate lordosis of the cervical and the lumbar sections of the spine, moderate kyphosis of the thoracic and the sacrococcygeal sections, a forward pelvic inclination of about 30°, neutral rotation of the femurs, and the head centered so that a plumbline dropped from the mastoid process passes through the middle of the shoulder and the hip and just anterior to the knee and the lateral malleolus of the ankle. Deviations from this basic position are due to (1) structural change of injury or disease, and (2) displacement of spine and pelvis caused by force of gravity overcoming muscle tone.

Importance of Posture

The orthopaedic surgeon is concerned with poor posture because of (1) a possible structural fault due to injury or disease, (2) muscle inadequacy and easy fatigue superimposed upon an inefficiently disposed skeletal framework, (3) frequent ligamentous strains and pains due to failure of muscular support, (4) abnormal bone growth in adaptation to the altered position, (5) cosmetic reasons and (6) the possible eventual development of a rigid, degenerative arthritic spine, particularly in the hyperextended cervical and lumbar segments. Improper spinal posture (accentuated anteroposterior curves, increased pelvic inclination) is eventually associated with a defect about the shoulders and the chest (scapulae forward placed, rhomboids weak, chest sunken) and about the lower extremities (flexion and internal rotation at the hips, knock-knees, flatfeet). Therefore, correction of the spinal and pelvic deformity has a beneficial effect upon these secondary deformities.

Pathologic Physiology

In the course of evolution from the quadruped to the orthograde animal, the relatively straight spine of the former develops forward and backward curves in the latter as it yields to the force of gravity. At the same time, the paraspinal and the gluteal muscles, which maintain the erect posture, become more fully developed.

[74] Michele, A A., and Krueger, F J.: Surgical approach to the vertebral body, J Bone & Joint Surg 31A:873, 1949.

[75] Burt, H A : Effects of faulty posture, Proc. Roy. Soc. Med. 43 187, 1950.

[76] Goldthwait, J. E., Brown, L. T, Swaim, L. T, and Kuhns, J. G.: Body Mechanics in Health and Disease, Philadelphia, 1934, 1937.

[77] Kuhns, J. B.· J. Bone & Joint Surg 24.547, 1942.

[78] Wiles, P : Lancet 1:911, 1937.

In the erect position, if the spine were a perfectly straight, balanced column of blocks, muscular support would be wholly unnecessary. However, forward and backward displacement of the vertebrae requires constant muscle control to maintain balance. This is obtained by continuous minimal muscular contractions, designated as postural tone. Constant postural tone depends upon proprioceptive sensory impulses (sense of position and stretch) from muscles and tendons, vestibule of the internal ear, and eyes to the central nervous system which reflexly stimulates the muscle. This mechanism effects the adjustments necessary to maintain balance as new movements and positions are assumed. In each muscle only a few fibers at a time contract, then others, the continual change of contracting fibers holding fatigue to a minimum. When the spine becomes displaced and unbalanced, more muscle power is needed to control the erect position, a greater number of muscle fibers are called into play at more frequent intervals, and fatigue develops earlier. Muscle insufficiency allows the spine to sag even further, and strain is thrown upon the ligaments with consequent pain Ordinarily, adequate musculature holds and moves the vertebrae in a mid-position so that ligaments are never under tension unless the extreme limit of the movement is reached

Persistence of an extreme lordosis with its unnatural stresses thrown upon the posterior articulating facets eventuates in later years in traumatic degenerative changes. The alterations are most marked at the point of greatest angulation, usually the lumbosacral junction.

An increased dorsal curve in the thoracic spine interferes with normal growth of the vertebral bodies. Anterior compression forces on the forward end of the growth plates results in wedging, especially at the lower thoracic spine. This suggests abnormal posture as a cause of Scheuermann's disease.

The Deformity

When abnormal increase of a curve occurs in a section of the spine, the curves above and below become accentuated in the opposite direction in order to maintain balance Thus, an increase of the thoracic kyphosis becomes associated with increased lordosis in the cervical and the lumbar areas. This is often described as the *lordotic back*. If lumbar compensation is insufficient, the thoracic portion of the trunk (and the mastoid plumbline) remains displaced posterior in relation to the pelvis, and the typical *swayback* is produced. Frequently, the *pelvis inclines forward* sharply in attempting to compensate, and an acute lumbosacral angle is associated. Hyperextension at the lumbosacral junction eventuates in a facet syndrome (q.v.). Forward pelvic inclination becomes a resistant deformity if allowed to persist. The hip flexors shorten, and the extensors (glutei) become lengthened and inefficient. Inadequacy of the glutei allows the thighs and the knees to rotate internally and additional deformities develop, such as knock-knees and flatfeet.

Wiles regards the posture of the hip joint as the key to that of the whole body because it determines (a) the pelvic inclination, the pelvis being the foundation for the spine, and (b) the rotation of the legs. The pelvic inclination can be measured by drawing a line from the upper border of the symphysis pubis to the posterior superior iliac spine. The normal is from 28° to 31°.

One may argue that the pelvic inclination is the starting point of the deformity. Actually, the pathogenesis, in the absence of bone injury or disease, is unknown. However, the position of the pelvis is a convenient starting point for determining the type of deformity and treatment.

Increased Pelvic Inclination

The following are the main features:

1. Ventral displacement of head with increased lordosis of neck
2. Round shoulders, flat chest
3. Increased dorsal kyphosis and lumbar lordosis
4. Forward pelvic tilt and prominent abdomen
5. Internal rotation of thighs, pronated feet, knock-knees

Decreased Pelvic Inclination

When the plane of the pelvis is disposed horizontally, balance is attained by the spine in one of two ways:

1. The spine straightens by reduction of cervical, thoracic and lumbar curves, and a *flat back* results. Such a back has a pro-

nounced tendency to stiffen later in life and cause intractible backaches.

2. The lumbar lordosis persists and displaces the upper trunk backward. Balance is regained by forward bending of the thoracic spine, creating a *round back.*

Postural Lateral Curvature

Lateral curvature of the spine is often discovered accidentally in adolescents, especially girls. The displacement includes the entire thoracic and lumbar spine and consists of a long, smooth C-curve, most often toward the left side. The deformity can be fully corrected by voluntary muscular effort and by bending forward, sitting, or standing on one leg. Rotation and structural deformity of the vertebrae never occur, and the postural curve corrects *spontaneously.*

Attempts to classify various types of postural deformity appear to be unnecessary. Treatment is directed at restoration to positions which are least injurious and provide maximum mechanical efficiency.

Etiology

Muscular inadequacy is the most probable cause of poor posture. Weak muscle tone may be inherited or follow a severe illness, malnutrition, or a prolonged period of inactivity. The factor of muscle insufficiency is most felt during periods of rapid growth when the muscles are elongated in an effort to keep pace with lengthening bony structures.

Contributing factors include nervous disorders, overwork, faulty school furniture, tight clothing, etc., all of which cause the child to sit or stand badly, thereby developing abnormal postural reflexes.

Treatment

The aims of treatment are restoration of balance, reduction of excessive curves, securing proper pelvic inclination and strengthen-

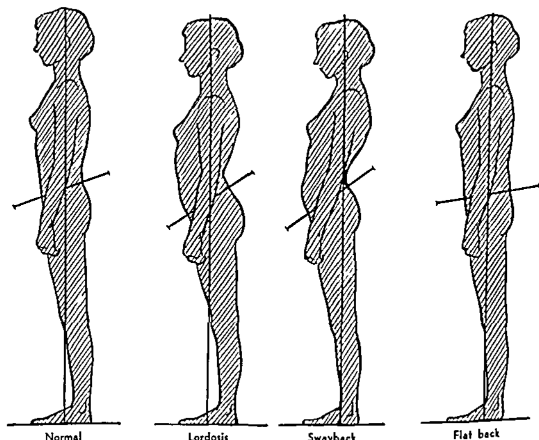


Fig. 514. Postural deformities. The dense oblique line indicated the degree of pelvic inclination as measured from the posterosuperior iliac spine to the upper border of the symphysis pubis. Restoring the normal pelvic inclination is the key to overcoming postural deformity. (Redrawn from Wiles, P.)



FIG. 515. Scoliosis due to shortened right lower extremity. Note the pelvic tilt. Correction obtained by balancing the lower extremities.

ing the musculature. This can be accomplished by:

1. **Mobilizing the Spine**

A. *Exercises.* These are designed to effect contraction of muscles which will reduce the deformity. Active contraction of the agonist, particularly when the movement is performed against resistance, is accompanied by reflex relaxation and lengthening of the antagonist.

The patient lies in the prone position, and restraining bands are placed across the lumbar area to restrict motion at this level. Extension of the thoracic spine is accomplished by slowly and repeatedly elevating the head and the shoulders from the table. Next, the patient lies in the recumbent position with the hips resting in a flexed position to avoid contraction of the hip flexors. The arms are folded across the front of the chest, and repeated "sit-ups" are performed Strengthening of the abdominal muscles effects flexion of the lumbar spine and elevates the front of the pelvis to the more horizontal position. Next, the glutei are exercised. The patient lies on the side and abducts against the resistance of the hand or gravity itself, then lies prone and similarly hyperextends the hip.

B. *Stretching.* Forceful manipulation should be avoided. It stretches muscles and excites the stretch reflex, which causes that muscle to contract more. It tears soft tissue, which results in scar contracture and rigidity Prolonged head halter and pelvic traction is more effective. General body exercises help to secure and maintain spine mobility.

2. **Postural Reflexes.** The patient must be conditioned to new habit postural patterns The patient is taught to assume a new position, and movements in various directions are made, each time returning to the starting new position. A constant consciousness of this position must be developed and held throughout the waking hours. By maintaining a stubborn voluntary control, the new postural reflexes are acquired. Exercises directed toward specific muscle groups aid voluntary control of the new position. The patient is taught to hold the head centered and erect, the shoulders held back, the abdomen flattened, the pelvis held at the proper level, and the knees facing forward.

3. **Constitutional Care.** Attention must be directed to adequate nutrition, correction of anemia and disease, avoidance of fatigue, and elimination of psychiatric factors.

SCOLIOSIS

Scoliosis, defined, is lateral deviation of the spine. It is important because it produces body disfigurement and predisposes to backache in adulthood. When deformity is extreme, the intra-thoracic and the intra-abdominal organs are displaced and even compressed, and their function is compromised; life expectancy may be reduced.

ETIOLOGY

The great preponderance of cases are idiopathic. However, intensive search for and correction of an underlying cause should be

attempted. The following is an outline of the known causes:[70]

I. **Congenital Scoliosis**
   A. Without manifest bone changes (idiopathic)
   B. With manifest bone changes
      1. *Hemivertebra.* A wedge-shaped half of a vertebral body is interposed between 2 normal vertebrae.
      2. *Maldeveloped vertebra.* One half of a vertebral body may be less developed than the other half, resulting in a wedge-shaped vertebra.
      3. *Fusion of irregularly developed vertebrae*
      4. *Spina bifida.* Especially in the cervical and the dorsal areas where severe curvatures may result
      5. *Lumbosacral malformations.* For example, asymmetric sacralization of the 5th lumbar vertebra

      Congenital lesions are usually multiple. Therefore, the vertebral lesions may be associated with Sprengel's deformity, webbed fingers, fusion of ribs, etc.

II. **Acquired Scoliosis**
   A. Idiopathic
   B. Definite causative condition
      1. *Poliomyelitis.* In paralysis, muscular imbalance allows the spine to displace to one side. The force of the unopposed muscles is operative, even in recumbency, but the deformity is worse and more progressive in the upright position when the force of gravity is added. The muscle involvement must be asymmetric to produce scoliosis. When paralysis is symmetric, regardless of the degree of involvement, a curvature does not eventuate. Asymmetric involvement of the abdominal muscles is as effective as involvement of the back muscles in producing a spinal curve. Paralysis and contracture of hip muscles and a tight iliotibial band will cause fixed pelvic obliquity and, secondarily, a lumbar curve Atrophy of bone in poliomyelitis may result in vertebral deformity. Reduction in rate of growth in one lower extremity involved by poliomyelitis produces inequality of limb length and consequent displacement of the spine in the standing position.
      2. *Other C.N.S. diseases.* Friedrich's ataxia, syringomyelia, spastic paralysis, neurofibromatosis, hysteria
      3. *Rickets.* Although today considered rare, related conditions as vitamin-resistant rickets, renal rickets and the adult osteomalacic type are observed occasionally. The vertebrae are osteoporotic, particularly at their centers. The disk spaces are widened, and the margins of the bodies are ill-defined.
      4. *Faulty posture*
      5. *Unequal lower-limb length.* Deformity disappears in the sitting position.
      6. *Visual or auditory disturbances*
      7. *Torticollis,* organic or functional
      8. *Thoracic disease or surgery.* (a) Unilateral cicatricial contracture and atelectasis; thoracic spine deviates to opposite side. (b) Thoracoplasty, weakening the thoracic cage; thoracic spine deviates to the ipsolateral side. The higher and the more proximal the level of rib resection, the greater the deformity.
      9. *Spinal disease.* The deformity is usually mild. Spondylitis deformans, fracture or dislocation, osteomalacia, dyschondroplasia, vertebral epiphysitis, syphilis, tuberculosis
      10. *Organic heart disease*
      11. *Heredity*
      12. *Miscellaneous*

The most common type by far is the idiopathic. The curve appears and progresses during the growth period, particularly in the preadolescent and adolescent states, and halts as growth ceases Girls, especially rapidly matured and precocious, are predisposed. This suggests an endocrine origin.

The following discussion is limited to the idiopathic type of scoliosis, but the clinical picture and treatment may be applied to other types, taking into account the variations due to respective operative factors.

[70] Kleinberg, S Scoliosis, Baltimore, Williams & Wilkins, 1951.

Pathogenesis

Normally, the main central portion of the vertebral body is ossified early, and the superior and the inferior surfaces are surmounted by an ossified rim known as the apophyseal ring. Between each apophyseal ring and the central ossification center exists cartilaginous tissue similar to the epiphyseal plate of long bones and responsible for the growth in length of the vertebra This epiphyseal tissue is subject to the same influences as in the long bones, namely compression, infection, traction, etc. Temporarily compressing the growth tissue, as by stapling, will arrest growth in length until the restricting force is removed. Unilateral stapling can be utilized to produce a scoliotic curve or to correct it. Risser utilizes the Hueter-Volkmann epiphyseal pressure rule in treatment By bending the spine against the curve, compression forces are greater on this side, and ipsolateral restriction of growth is effected. On the opposite side, spaces are created between the vertebrae, and traction is exerted; growth may continue here at the normal or increased rate, and bony substance is built up into the created spaces, thereby overcoming the wedging deformity. The same principle is used in overcoming the anterior wedging deformity in vertebral epiphysitis.

The changes which take place during active progression of the curve strongly suggest involvement of the epiphyses as the cause of scoliosis. The apophyseal rings become rarefied and fragmented. The disk spaces become mottled and indistinct, and the outline of the vertebral bodies becomes hazy The changes are most pronounced at the apex of the curve. As the disease process regresses, these structures become more distinct, although altered in shape. It is not altogether inconceivable that infection, deficient circulation, or endocrine influence reduce resistance to pressure forces, and asymmetry results. Very commonly, scoliosis has its origin before the epiphyseal plates ossify, and changes are not demonstrable in roentgenograms

The actual mechanism in the production of scoliosis is unknown. The following are the known factors. The spine is a flexible column, supporting the head and the trunk and permitting motion in various directions. Forward flexion and extension occur chiefly in the lumbar and the lower dorsal spine. Side-bending is accompanied by a rotatory motion. The combined movement takes place in the lumbar region, and the vertebral bodies usually rotate toward the concavity of the curve. However, if the body is in the forward flexed position, side-bending takes place chiefly in the dorsal region, and rotation of the vertebral bodies is usually toward the convexity of the curve.

Viewed from the front, the body is aligned so that the head is centered over the middle of the sacrum. Seen from the side, a plumbline (or center of gravity) passes through the ear, the shoulder, the greater trochanter, and the lateral malleolus of the ankle. Co-ordinated muscle action maintains this erect position or attempts to maintain body balance. If, for example, the dorsal spine becomes rounded and displaced backward by vertebral epiphysitis, balance is secured by increase of cervical and lumbar lordosis. If the lumbar spine is displaced laterally because of a shortened lower extremity, the thoracic spine in compensation displaces to the opposite side, thus restoring balance. The development of compensatory curves is an attempt on the part of the body to restore the main body weight to the center of gravity.

Deviation of the spine to one side imposes greater force on the concave side of the vertebrae. The resultant wedging of the vertebral bodies encourages further displacement, and a vicious cycle is established.

Descriptive Terms

The side toward which the convexity of a curve is directed is designated as "right" or "left." The segment of the spine is named "dorsal," "lumbar," "cervical," "cervicodorsal" and "dorsolumbar." When deviation of the spine occurs in one direction only, the terms "C-curve," "simple curve," or "total curve" are used interchangeably. "S-curves" or "compound curves" describe displacements in both the right and the left directions. The "primary curve" is that which appears first. A "secondary" or "compensatory curve" is that which develops as a compensating balancing curve to the primary one. "Mild," "moderate," or "severe" describes the degree



FIG. 516. Postmortem specimen, displaying extreme intrathoracic deformity and compression.

of involvement not only in reference to the amount of lateral displacement but also to the amount of associated deformities, rotation and soft-tissue contracture. "Functional scoliosis" is a term applied to a curve not associated with deformity and soft-tissue contracture, one which can be corrected voluntarily. A better designation is "nonrigid scoliosis," because it implies that maximal correctibility is possible. However, when the nonrigid curve is allowed to persist, deformities of the vertebrae and the ribs and contracture of soft-tissue eventuates in the "rigid" or "structural curve."

PATHOLOGY

The typical nonrigid curve occurs as a single C-curve in the dorsal and the lumbar areas, usually to the left. As viewed from behind, the body appears asymmetric. The ribs on the concave side are closer together, and the lower border of the thoracic cage on this side approximates to the ilium The disk spaces are widened on the convex side and narrowed on the concave No actual bony deformity is noted. The absence of adaptive soft-tissue contractures is evidenced by ability of the spine to move through its full range of motion in all directions. Progression of the curve is variable. It may persist as a mild nonrigid type. Or, it may proceed to become greater in degree and rigid. Secondary balancing compensatory curves usually develop above and below the initial curve and, because they are of shorter duration than the primary curve, they are much less rigid and show little or no deformity. All varieties and combinations of curves are possible. As a general rule, spinal growth ceases coincidental with completion of growth of the iliac apophysis Beyond this time no further progression of the spinal curve is possible.

When deviation of the spine progresses and becomes more persistent, the vertebrae rotate and become more wedged. The vertebral bodies usually turn toward the convex side, and the spinous processes in the opposite direction. The thoracic vertebrae in rotation displace the ribs backward on one side posteriorly and forward on the opposite side anteriorly. Clinically, this is manifest as posterior and anterior protrusions of the thoracic cage, well revealed in the forward bent position. On the convex side the intercostal spaces are widened, whereas they are narrowed on the concave side. All soft-tissue structures are

adaptively contracted and thickened on the concave side and stretched on the convex side. The intervertebral disks are wedged on the concave side, while the nucleus pulposus is displaced toward the opposite side. All structural changes are most pronounced at the apex of the curve. The sternum is not deformed but is displaced about its lateral or transverse axis. The degree of deformity is generally proportionate to the duration of the curve, so that the first-appearing, or primary, curve displays the greatest change and the secondary, or compensatory, curve the least change.

The presence of structural changes implies passage of sufficient time to allow development of compensatory curves, so that S-curves are found almost invariably. A curve with structural changes and rigidity is one which is unchanged by position, traction, or voluntary effort. The rotation of the vertebral bodies in the thoracic area is usually toward the convexity of the curve, and the ribs on this side are carried backward and become angulated The rib deformity also occurs anteriorly on the contralateral side. The posterior angulations may become so extreme as to form a vertical ridge known as a "razor back," which is best seen with the patient bent forward. It is an indication of the degree of vertebral rotation. The intrathoracic structures on the side of the curve convexity are compressed, and the lung may be atelectatic. Vital capacity is reduced. The heart is displaced downward, and intrapulmonary obstruction results in right-sided hypertrophy and dilatation. The aorta is displaced In the lumbar region, the bodies again rotate to the convex side as a rule, and the transverse processes on this side also turn backward and cause the soft-tissue structures to protrude, thereby creating a deformity. This, too, is more visible in the stooped position The abdominal organs are displaced downward toward the pelvis. Rarely is the spinal cord compressed except in severe scoliosis Usually, the sacrum is not involved. Rarely, it may form part of the lumbar curve but is never deformed.

Associated pathology contributing to formation of a curve includes single or multiple hemivertebrae, extra ribs, resection of the bases of ribs, etc.

Symptoms

Subjective complaints are absent or minimal in childhood. Backache, back stiffness and back weakness are more likely with advancing age, particularly after heavy and prolonged activity.

Clinical Findings

Deformity of the spine in the lateral direction may occur without change in bodily contour because of balancing curves. The defect is found early by a routine examination in childhood when the physician focuses his attention in this direction. The unbalanced curve is more generally noted early. In the erect position the body is displaced to one side. Posteriorly, on the ipsolateral side, the ribs protrude backward, displacing the scapula. This deformity is rendered more prominent when the patient bends forward at the hips. Also on this side the shoulder is elevated, and the loin fold is obliterated. The opposite shoulder is lowered, and the loin fold is deepened. A chest protrusion also occurs anteriorly on the contralateral side but is milder than the posterior protrusion. The accentuated loin fold renders the hip more prominent, and the lower costal margin approaches the iliac crest.

In the presence of a shortened lower extremity, the iliac crest on this side is lower, and the main curve is in the lumbar area. In the sitting position, the pelvis is level, and the lumbar curve is lessened.

The flexibility of the curves varies When no rigidity is present, the curve straightens and even bends in the opposite direction as the body bends toward the side of the convexity. Restriction of correction of the curve depends on the degree of structural change and rigidity. The mobile curve will straighten spontaneously in recumbency.

Examination

A history of the complaint should include a thorough search for causative factors, as poliomyelitis and a shortened lower extremity. The factor of heredity should be ascertained. It is exceedingly important to determine the rapidity of appearance and progression of the deformity.

Recorded complaints include prominent



Fig. 516. Post-mortem specimen, displaying extreme intrathoracic deformity and compression.

of involvement not only in reference to the amount of lateral displacement but also to the amount of associated deformities, rotation and soft-tissue contracture. "Functional scoliosis" is a term applied to a curve not associated with deformity and soft-tissue contracture, one which can be corrected voluntarily A better designation is "nonrigid scoliosis," because it implies that maximal correctibility is possible However, when the nonrigid curve is allowed to persist, deformities of the vertebrae and the ribs and contracture of soft-tissue eventuates in the "rigid" or "structural curve."

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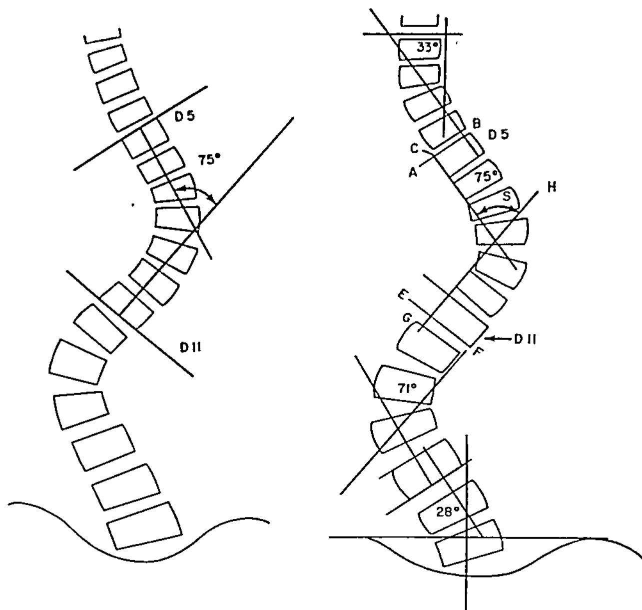


FIG. 518. (*Left*) Measurement of the angle of a curve.

1. Locate the top vertebra (D5 in this case). The top vertebra of the curve is the highest one whose superior surface tilts to the side of the concavity of the curve to be measured. (The superior surface of the vertebra above it usually tilts in the opposite direction, to the side of the convexity, but may be parallel. The intervertebral space on the concave side is usually wider above the top vertebra and narrower below it, but if there is vertebral wedging, this I.V. space may vary.)

2. Locate the bottom vertebra (D11 in this case). The bottom vertebra is the lowest one whose inferior surface tilts to the side of the concavity of the curve to be measured. (The inferior surface of the vertebra below it usually tilts in the opposite direction, to the side of the convexity, but may be parallel. The I.V. space on the concave side is usually wider below the bottom vertebra and narrower above it, but if there is vertebral wedging this I.V. space may vary)

3. Erect intersecting perpendiculars from the superior surface of the top and the inferior surface of the bottom vertebrae of the curve.

4. The angle formed by these perpendiculars is the "angle of the curve."

(*Right*) Drawing from a roentgenogram showing measurement of all 4 curves of figure at left. (Note major structural factors, wedging, rotation and position of curves, and good compensation.)

Left angular tilts 75° and 28° = 103°. Right angular tilts 33° and 71° = 104°. (From Cobb)



FIG. 517. Demonstrating mobility and correctibility of curves. L.G Onset of scoliosis at about 12. Surgical treatment at age 13 Preoperatively, mobilization of dorsal spine effected by suspension in a hammock. Roentgenograms taken while side-bending demonstrate straightening of the dorsal spine, which is almost comparable with the secondary lumbar curve

hip, sunken waistline, protruding ribs and shoulder blade, elevated shoulder, dropped shoulder, prominent breast, poor posture, backache and fatigue, dsypnea and root pains

The patient should be completely undressed. The height, the weight, the general nutrition, and the condition of the heart and the lungs should be noted. With the patient standing, the trunk is inspected from the posterior aspect, and a plumbline is dropped between the gluteal cleft. Asymmetry of the body, as for example in a single left C curve, shows the following: (1) the left shoulder is higher than the right and is carried forward; (2) infolding of the right loin and flattening of the left; (3) prominent right iliac crest; (4) thoracic cage shifted to the left; (5) the left side of the chest is more prominent than the right; (6) the head is to the left of the center of gravity. The patient then bends forward to a right angle at the hips, and the back protrusions are well observed. This indicates the degree of rotation and fixation of the curve. The spinous processes are marked with ink. Then with the patient erect, he is asked to bend to each side, and the amount of straightening of the curve is recorded Next, the patient sits on a hard flat surface, and any spontaneous correction of the lumbar curve is noted. The buttock on the side of lumbar convexity is elevated on a block, and the amount of correctibility of this curve is estimated. An additional measure of spine straightening can be obtained by suspending the body by an overhead head halter and noting the line of spinous processes. Then the patient lies in the prone position, and spontaneous correction of the curve is determined.

Photographic records are essential at this time. The bodily asymmetry and protrusions can be compared with pictorial records of the final result.

Roentgenographic Examination. X-ray films constitute the most important phase of the examination. By them one is able to ascertain with great accuracy the extent, the degree and the mobility of the curves. Associated deformities are revealed. Progression of the curve can be followed. With the patient in the standing position, an antero-posterior view of the dorsal and cervical spine is taken, and another of the lumbar spine and the sacrum. The two views are pieced together so as to form a composite picture. The curves, the vertebral contour and rotation and associated congenital deformities, if any, are visualized. The angle of each individual curve is measured and recorded. When the body is symmetric and the head is centered over the mid-line, a curve in one direction is balanced by spinal deviation of the same degree in the opposite direction. Similarly, when multiple curves exist, the sum total of degrees of curves to the right should be equal to the total degrees of curves to the left. When a primary curve is not compensated for by another curve or when there are several curves whose total deviation is less than the primary curve, the body is unbalanced and asymmetry re-



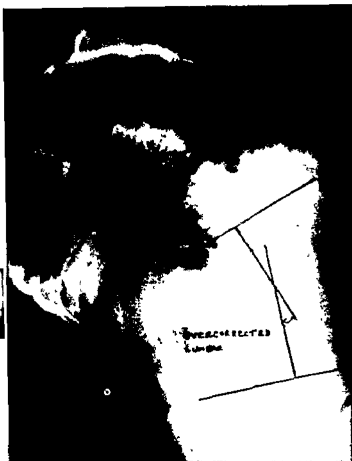


Fig. 520. (*Left*) Reduction of thoracic curve by bending to the right. Residual arc indicated by arrow. (*Right*) Bending film, showing reduction of lumbar curve. The overcorrection indicates excellent mobility. Full spontaneous correction may be expected.

length of the spine terminates at a time coinciding with completion of development of the iliac apophysis ossification center. The latter process is seen in roentgenograms as a dense line at the iliac crest which develops from the anterior toward the posterior iliac spine and terminates when it curves inferiorly at its posterior extremity.

Measurement of the Curve The top and the bottom vertebrae are identified. The intervertebral spaces in a curve are wider on the

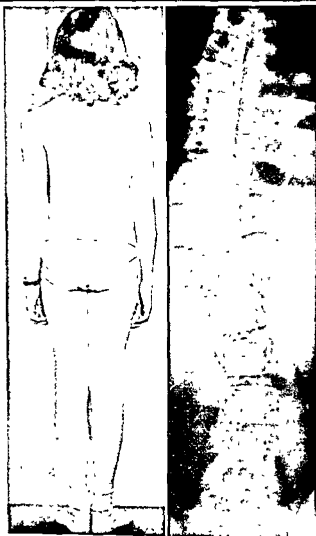


Fig. 521. D.H. (*Left*) Clinically marked improvement. The bodily contour is well balanced Prominence of the scapula and the dorsal chest cage is permanent. (*Right*) Final correction. The small residual curve in the thoracic is balanced by a similar one in the lumbar spine. The bone graft is well seen on the concave side of the thoracic curve.





FIG. 519. D.H. (*Left*) Standing, elevation of right shoulder and prominent scapula well shown. (*Center*) Bending forward. The dorsal chest protrusion is seen best in this position. (*Right*) Standing anteroposterior view of spine from T1 to sacrum. Typical primary thoracic and secondary lumbar curves. The actual degree of each arc is shown by the double-headed arrow.

sults. The compensatory curves may occur above or below the primary curve.

Next, films are taken to determine the amount of mobility and correctibility of each curve. One composite is made with the body bent lateralward against the convexity of the thoracic curve, and another when bent against the convexity of the lumbar curve. In each instance the convexity will straighten to a degree corresponding to the amount of residual mobility. The curves are measured and recorded. An anteroposterior projection view is taken in the recumbent position to measure the amount of spontaneous correction.

It is essential to identify the *primary curve*. This is the earliest-appearing spinal deviation, and other curves are *secondary* and develop to compensate for the primary curve. The latter result as an effort of the body to regain the erect position and to re-establish the head over the center of gravity. When the primary curve decreases, the tendency is for the secondary curves to lessen likewise. A curve of long standing tends to develop structural changes, and the vertebrae rotate, resulting in rigidity and incorrectibility of this portion of the spine. Therefore, as a general rule, the primary curve exhibits these findings in greater degree than the secondary curves, which are of shorter duration. Occasionally, difficulty is encountered when all curves are of long duration and display deformity and rotation which are apparently uniform. Ferguson[80] uses the following rules to identify the primary curve: (1) the middle one of three curves is usually primary; (2) the greater curve is the primary; (3) the least correctible curve is the primary; for example, in the previously described pelvic tilt x-ray film, if the lumbar curve fails to correct, it is a primary curve, whereas, if it straightens, it is compensatory. Normally, the dorsal spine is less flexible than the lumbar, and the element of rigidity must be interpreted in the light of this knowledge.

Risser has demonstrated that growth in

[80] Ferguson, A. B : The study and treatment of scoliosis, South. M. J. 23 116, 1930

a moderate degree is more probable when the age of onset is in early childhood.

Thoracic. Predominantly in females and to the right, usually it is first noticed at about age 10 and matures at 16. It generally includes 6 vertebrae from T6 to T11 with the apex at T8 or T9, and the rotation is extreme. Small countercurves exist above and below. The thoracic curve progresses more rapidly and produces more deformity than any other curve. It is also less reversible and therefore should be handled early and aggressively. Cases of early onset, before the age of 10, frequently reach angles greater than 100°. However, extensive deformity is the great danger no matter what the age of onset.

Cervicothoracic. This is uncommon, usually first noticed at 15, is never pronounced, occurs mainly in girls and is convex to the left. The apex is at T3 and includes 4 to 6 vertebrae from C7 to T5 which are wedge-shaped and markedly rotated. Below this exists a long, low thoracic countercurve. The shoulder is slightly elevated on the side of the convexity, but the general body alignment is good, and intervention is not indicated.

The Main Determining Factors in Prognosis

1. **Pattern of Curve.** Progression to deformity is proportionate to the predominance of thoracic over other components.

2. **Age of Onset.** The earlier the age of onset, the greater the tendency to deformity. Thoracic curves usually appear earlier.

3. **Alterations About the Apex of a Curve.** The important prognostic signs seen in roentgenograms indicating active progression of the curve are: (1) vertebrae adjacent to the apex are osteoporotic and hazily outlined; (2) intervertebral spaces are irregularly outlined and narrow; (3) a translatory shift in the transition vertebrae between the thoracic and the lumbar curves indicative of ligamentous relaxation; and (4) marked wedging at the apex. These changes are more pronounced in proportion to rapidity of progression of the curve.

Treatment

Prophylactic treatment is useless in the idiopathic type of scoliosis. Where causative factors can be identified, these should be removed. The following are important in that proper treatment will prevent or reduce spinal deformity:

Causative Factors

1. *Poliomyelitis.* Recumbency should be continued until maximum recovery is attained. Asymmetric muscle involvement of the back or the abdomen, pelvic tilt associated with a contracted iliotibial band, and lower limb shortening are causes of spinal curvature. One must be aware that a spine may be straight in the erect position because of soft-tissue rigidity. The spine becomes slowly deformed. Close observation is necessary when the paralytic patient assumes the upright position. If recumbency is contraindicated, a rigid corset or brace should be worn. Other neurogenic types of scoliosis are treated similarly.

2. *Rickets.* This type tends to become severe but fortunately it is rare. Recumbency, diet, vitamins, ultraviolet rays and the use of a plaster bed are effective.

3. *Posture.* Correction of poor posture theoretically eliminates unequal pressure on the vertebral epiphyses. The role of this factor is greatly disputed.

4. *Physical and Mechanical Causes.* Treatment is directed toward correction of limb inequality, impaired vision and hearing and torticollis. Because a healed intrapleural infection and its resultant scarring can cause scoliosis, trunk exercises, breathing exercises, brace supports, and general gymnastics are prescribed. In thoracic surgery, one must avoid removal of the posterior ends of the ribs.

Idiopathic Type

The active treatment of idiopathic scoliosis varies in different clinics only insofar as the use of exercises is concerned. Kleinberg states that increase of deformity can be prevented in about 75 per cent of cases by persistent and long-continued exercises, and in about 50 per cent of these the deformity can actually be reduced. This opinion is shared by many. However, a study by the American Orthopedic Association in 1941 resulted in conclusions that (1) patients treated by exercises either had no reduction of the deformity or the curve was worse; (2) when supports are used for correction, without fusion, complete loss of correction is the rule; (3) correction by the turnbuckle jacket and fusion produced the highest percentage of good results.

Indications for Surgical Treatment. A

side of the convexity and narrower on the opposite side. The spaces immediately above or below the curve are either of equal width throughout or are wider on the side of the concavity. The relation of the spinous processes to the body of the vertebrae determines the rotation Rotation is greatest at the apex of the curve and diminishes toward each end where the nonrotated or neutral vertebra is found. A line is drawn parallel with the upper and the lower surfaces of the top and the bottom vertebra, respectively. A perpendicular is drawn from each of these transverse lines, and an angle is formed where these two perpendiculars cross. This is the angle of the curve.

Prognosis in Scoliosis

Before one can decide on whether to employ conservative or surgical treatment, it is important to know the natural course of a curve and its probable outcome. The curve can begin at any age in childhood and stop progressing any time before growth is completed. The end of growth is shortly after the completion of formation of the iliac apophysis, and this may be used as a good sign to determine the end of progression of the curve. A curve which will not increase is designated as "mature." A curve may increase slowly, then remain stationary and suddenly increase very rapidly. Therefore, a case should be followed for a long time, even for years, although it appears that the curve is unchanged. The patient is studied by roentgenograms taken with the patient in the standing and the recumbent positions, and the angles of the curves are measured and recorded. Side-bending films will determine the increase of rigidity (incorrectibility) of the curve. Clinical examination is done to investigate deformity and mobility. The procedure is repeated at regular intervals, usually at 3-month periods, or sooner if one suspects a rapidly changing situation. Surgical intervention is undertaken when severely incapacitating and deforming curves are anticipated and before they become rigid A curve is more likely to be serious if it starts early in childhood and if its main component is the thoracic portion of the spine.[81, 82]

[81] James, J I P. Idiopathic scoliosis, J Bone & Joint Surg. 36B 36, 1954.

[82] Ponseti, I V., and Friedman, B · Prognosis in idiopathic scoliosis, J. Bone & Joint Surg. 32A 381, 1950.

Typical Curve Patterns and Their Characteristics

Lumbar. Benign, predominantly in females at about 12 years of age; matures at about 15 It generally includes 5 vertebrae from T11 to L3 with an apex at L1 or L2. Two or 3 thoracic immediately above and the last 2 lumbar vertebrae form small balancing counter curves. The rotation of the spinous processes toward the concave side in the lumbar curve is also present in the counter curves to the same side but to a lesser degree. In general, this curve is minimal, nondeforming and asymptomatic It may be a cause of low back pain in adult life.

Thoracolumbar. Predominantly in females, it is first noticed at about 14 years of age and matures at about 16. It is usually to the right and includes 6 to 8 vertebrae from about T6 to L2 with the apex at T11 or T12. When the apex is at T11, the curve is usually better developed than when at T12. The counter curves are also greater in degree in the former than in the latter. The final curve is slight to moderate and hardly ever exceeds 40°. The cosmetic appearance of dropped shoulder and prominent hip is so mild that fusion is not required. Because of the lumbar element of this type, backache in later adult life is a possibility.

Combined Lumbar and Thoracic. This is the most frequent form. It occurs predominantly in girls at about age 12 and matures at 15. It is composed of two main curves. The thoracic curve, generally to the right, extends over 5 vertebrae from T6 to T10, with the apex at T7 or T8. The lumbar curve is in the opposite direction and includes 5 vertebrae from T11 to L4, with the apex at L2. Except for a slightly increased degree of the thoracic component, both curves are identical in the amount of rotation toward the convexity and the rate of progression In about 25 per cent of cases, the thoracic curve is greater, the apex lower, and the lumbar curve, being shorter, cannot compensate. Therefore, the body overhangs to one side. However, an upper cervicothoracic countercurve helps to align the body. The other 75 per cent of cases are not too deforming, and the general body appearance is satisfactory because the lumbar and the thoracic curves are identical and balance each other Progression of the curves to more than

the extreme of forward flexion, the patient grasps both knees and pulls them toward the abdomen, thus forcibly gaining further flexion This maneuver stretches the lumbosacral ligaments and reduces the lumbosacral angle and hence the lumbar lordosis. The patient then lies on his side. The head and the shoulders are raised upward. Then the uppermost extremity is abducted. These exercises are repeated on the other side. Next, in the erect position, bending exercises are performed, including forward bending, backward bending, bending to either side, and rotatory movements. Finally, the patient puts his hand over the side of the chest corresponding to the thoracic convexity and bends the head and the shoulders toward that side. These exercises designed to reduce the convexity and strengthen the musculature on that side are known as asymmetric exercises Other general exercises may be added as necessary. Both types are done daily. They are initiated with a bare minimum of effort and are gradually increased in force and duration. Fatigue is avoided.

STRETCHING. The daily application of longitudinal and lateral traction forces will mobilize the spine effectively and gradually. The patient lies in the recumbent position. A head halter with about 10 pounds of weight attached pulls proximally. A pelvic girdle and traction straps with about 20 or 30 pounds of weight pulls distally. This type of traction is better tolerated by the patient than bilateral leg traction and may be removed at intervals for exercises. A sling is placed about the apex of the curve, and 5 pounds of weight exerts a pull laterally toward the concavity. This weight is gradually increased until maximum tolerance is reached. Other lateral slings may be added above and below this first

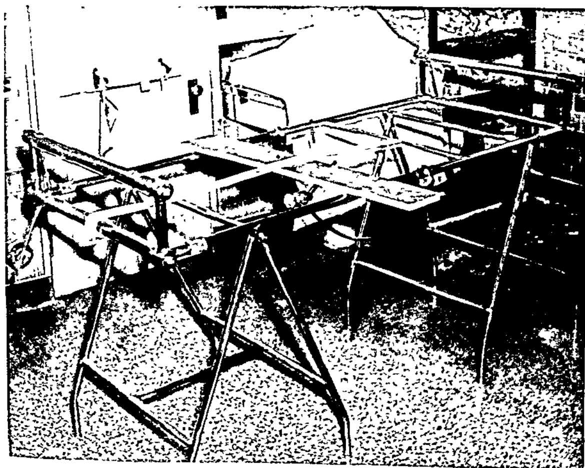


FIG. 523. The Risser localizer table. The localizer arc is suspended from the side bars of the frame, and the localizer pivot-point jack is fastened on the arc. Ratchets at the head and the foot of the frame are for parallel traction. A canvas strap extends from the pull-out bar to the ratchet bar at the foot of the frame.

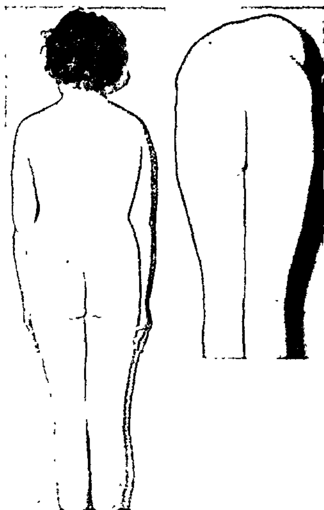


FIG. 522. Scoliosis, moderate, almost compensated. Progressive increase, particularly of the chest deformity, was an indication for a spinal fusion. The extent of the posterior chest protrusion on the right side is well shown in the forward bent position.

curve which is mild or curves which are large but well-balanced and therefore nondeforming are in themselves no indication for surgical *intervention. In the great majority of cases,* spinal fusion after preliminary measures to mobilize and reduce the curve is effective in diminishing the deformity and preventing it from becoming worse. Fusion of the spine in childhood will not interfere with growth in length of the spine. Deliberation and conservative treatment are hazardous, because a mild curve can rapidly become severe Reduction of deformity must not only be obtained but must be maintained The following are the definite indications for surgical intervention:

1. *A progressive curve*
2. *Deformity of the trunk,* regardless of whether or not spinal growth has ceased
3. *Pain,* particularly in older patients
4. *Hereditary history of severe scoliosis*

Thoracic curves are most likely to become severe and should be kept under close scrutiny. This is particularly true in primary cervicodorsal and high dorsal curves.

Presurgical Treatment. First, the spine should be thoroughly mobilized by stretching the soft tissue structures on the concave side of the curve and attempting to maintain the correction. An outline of this program as follows has given excellent results:

OUTLINE OF THE PROGRAM

A. *Mobilization Stage*
   1. Exercises
      (a) Symmetric
      (b) Asymmetric. Muscles on convex side strengthened
   2. Stretching
      (a) Longitudinal traction. Head halter and pelvic
      (b) Lateral traction. Sling over apex of curve
      (c) Overhead suspension of arm and leg on convex side

B. *Forcible Correction Stage*
   1. Risser turnbuckle hinge jacket
   2. Compression by a jack on Risser casting table (localizer cast)
   3. Corrective plaster jacket applied while lateral force is exerted

GYMNASTIC EXERCISES. Symmetric exercises are first done to develop the general bodily musculature and tone and are essential to retaining good posture. The patient lies in the prone position, clasps both hands behind the head and raises the head and the shoulders. Next, the lower extremities are raised with the knees fully extended. This contracts the gluteus maximus and the back extensors. Finally, head and shoulders and the lower extremities are raised simultaneously. The patient then assumes the recumbent position, folds both arms in front of the chest, and the head and the shoulders are raised. This contracts the abdominals and the hip flexors As the abdominals strengthen, this exercise is performed without the aid of the hip flexors by keeping the hips and the knees bent. At

The cast is allowed to dry for 2 days. Then an elliptical section of cast is removed on the convex side to allow space for bending. The edges of this window should be well away from the skin and protected by sufficient padding, because these edges will move toward the chest wall as the cast bends.

On the opposite side, a transverse cut is made in the cast and a turnbuckle is attached. This turnbuckle is turned until gentle resistance is encountered, and further separation is accomplished by adding a few turns daily.

One must be constantly on the alert for pressure sores developing and beginning brachial plexus traction neuritis, and replacement of the cast may be necessary. Roentgenographic study will determine when desired correction has been reached. Then the open area in the cast is filled in with plaster and firm wood struts, and the turnbuckle is removed.

Further fixation of the cast is secured by extending a strut from the headpiece to the thighpiece. Before surgery a large window is removed from the back sufficient to permit surgical asepsis and exposure. The underlying padding is fixed to the edges with adhesive tape.

THE LOCALIZER BODY CAST.[83] This cast was devised by Risser to overcome the chest deformity and to permit ambulation during the prolonged immobilization. By this method localized pressure is exerted posterolaterally over the rib angulation in the presence of head and pelvic traction. This localized force, directed at the rib angulation, forces the apex of the curve under the ends of the curve, thereby producing correction.

The patient lies recumbent upon a removable hammock strap suspended on a rectangular frame. The head is suspended on an overhead sling, the neck resting upon a pull-out bar. The lower extremities are flexed over a pillow, thereby obliterating the lumbar lordosis. A firm plaster mold is applied about the pelvis with the bony prominences well padded. Pressure against the lower abdomen is applied by a triangular block while the plaster is hardening. Then a pelvic belt is applied over the pelvic mold for traction

[83] Risser, J C: The application of body casts for the correction of scoliosis, Am. Acad Orthop Surgeons, Lect 12.255, 1955.

Next, head-halter traction is applied, and plaster is molded about the head from chin to occiput. The third step consists of applying pressure against the rib angulation by a localizer jack. The tip of the jack should be well padded. The suspension sling is removed from beneath the head and placed beneath the pelvis. Then the hammock is removed. Finally, the headpiece and pelvic portion are connected with plaster. It is important not to constrict the lower ribs. Finally, the cast is trimmed. A "V" cut is made from the suprasternal notch to the sides of the mandible, leaving the point of the chin free. To prevent pressure sores, a long linear saw cut is made across the full extent of the rib angulation. This is the typical cast for the long "C" curve.

When the curve is localized to the lumbar spine, the pelvis by unilateral traction is tipped sidewise against the convexity, and the localizer pad is directed laterally toward the transverse processes a little above the apex of the curve. A lateral pull is exerted by a lateral sling about the pelvic cast. This method not only corrects lumbar curves but also obliterates pelvic obliquity.

In the case of double curves if the major curve is in the thoracic region, pelvic pull is applied unilaterally upon the side of the concavity of the lumbar curve.

When curves are of long standing and are severe and rigid, daily localizer pressure may be utilized to mobilize the spine before the final preoperative correction and casting are done.

After the cast has been applied, a window is cut in the back of the cast to expose the surgical area. From 7 to 10 days after surgery the skin sutures are removed, and a new body cast is applied. Cast changes are continued every 3 or 4 months.

This method is also superior for applying antigravity casts for correcting and immobilizing anteroposterior curves, e.g., in Scheuermann's disease.

CORRECTIVE JACKET APPLIED WITH LATERAL TRACTION. The patient stands erect with the head suspended by overhead head-halter traction. A canvas belt encircling the body exerts traction laterally over the apex; countertraction is applied by lateral bands above and below the first band. A large felt pad is applied over the concave side for fur-



Fig 524. Risser wedging jacket for scoliosis.

sling to afford countertraction or to correct compensatory curves. Fishnet traction is a valuable addition. The patient is placed on his side in a fishnet suspended from the ceiling. The hand and the leg on the convex side are attached to ropes from above. Then the fishnet is lowered, and the sagging weight of the body acts as a passive corrective force This procedure may be repeated daily.



Fig. 525. Illustrating straightening of the dorsal spine in the Risser jacket. Spinal fusion performed in 2 stages.

Forcible Correction. The Risser turnbuckle hinge jacket is an effective contrivance developed for forcible reduction of a curve and is most suited for use in conjunction with spinal fusion. It is applied by placing the patient in the supine position on the casting table. The hips and the knees are flexed only enough for flattening the lumbar spine. A sling made of bandage is placed about the head and is used to displace the head toward the concave side, i e., in line with the upper limb of the thoracic curve. This is necessary because as the curve is straightened the head will not be extremely displaced toward the opposite side and unnecessarily stretch the brachial plexus. Stockinette is applied about the body and the thigh on the convex side. Sponge-rubber padding is fixed over bony prominences and over the sites of pressure caused by bending of the cast. Special attention is given to padding the chest on the lateral side of the convexity. The dorsal and the ventral prominences should be protected. Several layers of sheet wadding are snugly applied. Then plaster is rolled on and should include the chin and the thigh on the side of convexity. If the dorsal curve is high, the entire head is incorporated in the cast. Hinges are placed on the front and the back over the apex of the curve and in a slightly eccentric position on the convex side so that distraction of the spine will occur during correction.

sion in severe scoliosis when paralysis threatens. Removal of a hemivertebra has been attempted in the lumbar region, but the operation is fraught with great danger, and the results are poor. The use of epiphyseodesis to retard longitudinal growth on the convex side is still in the experimental stage.

Scoliosis, even if well compensated, may be productive of painful degenerative arthritis in the adult. Spine fusion is indicated.

I

THE HOSPITAL FOR SPECIAL SURGERY

SCOLIOSIS HISTORY

Name Date History Number

Date of Birth Age Sex

Complaint: (High shoulder, prominent shoulder blade, high or prominent hip, prominent chest or breast, short leg, curvature, poor posture, awkward walk, pain in back, general fatigue, etc.)

Present History: Which deformity noted first

Age first noted by whom (patient, parents, doctor, teacher, nurse)
Age first treated what treatment (corset, brace, frame, exercises, traction)
Duration of treatment
Progress of deformity (stationary, increasing, decreasing, rapid, slow)

Family History: Give history of scoliosis or other deformity in each case. (other side)

Maternal grandmother (descent..............) Paternal grandmother (descent..............)
" grandfather (descent..............) " grandfather (descent..............)

		Age	standing height		sitting height	
Father	Age					
Mother	Age		"	"	"	"
Brothers	1	Age	"	"	"	"
(names)	2	"	"	"	"	"
	3	"	"	"	"	"
	4	"	"	"	"	"
Sisters	1	"	"	"	"	"
(names)	2	"	"	"	"	"
	3	"	"	"	"	"
	4	"	"	"	"	"

Past History: Birth difficult or normal. Evidence of trauma?

General health (robust, weak, normal weight, underweight, overweight, good color, pale, sickly, frequent illness) Illness—(age and duration) scarlet fever , measles , mumps chicken pox , whooping cough , polio , rickets, diphtheria, empyema , pneumonia , torticollis , Friedreich's ataxia chorea , tonsillitis menses, age onset, (regular, irregular, scanty, profuse, duration)

Examination Date

General posture (good, fair, poor). General development (good, fair, poor). Musculature (good, fair, poor). Nutrition (good, fair, poor) High shoulder (rt, left) Prominent scapula (rt, left) Prominent low ribs (rt., left) Exaggerated flank crease (rt, left). Prominent hip (rt, left). High hip (rt., left) List (rt, left). R. A L. A Rotation (no, mild, moderate, severe)
Standing height Sitting height Weight
Flexibility of spine (flexible, moderately fixed, fixed)
Curve (Note muscle imbalance on other side of sheet)
Curve corrected on forward bending? (No, slight, moderate, complete)
Curve corrected on suspension? (No, slight, moderate, complete)

Diagnosis—Scoliosis (check which)

- A Functional
- B Structural
  - I Osteopathic
    - 1 Congenital
    - 2 Thoracogenic
    - 3 Other osteopathic
  - II. Neuropathic
    - 1 Congenital
    - 2 Post polio
    - 3 Other neuropathic
  - III. Myopathic
    - 1. Congenital
    - 2 Muscular dystrophy
    - 3 Other myopathic
  - IV. Idiopathic

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ther correction. After applying the body cast, a large window is cut out over the concave side, and a small window is cut out over the convexity. Corrective pressure pads are inserted about once weekly through the latter window until the hollow side fills out. These casts are reapplied until maximum correction has been obtained.

A SEMIBENT JACKET is one applied in two separate sections, one about the chest and the other about the pelvis. This method is somewhat similar to the previously described Risser turnbuckle jacket. Correction is effected by bending these sections and filling in the interval with plaster

THE TRANSECTION JACKET is used particularly for a curve at the mid-section of the body. A complete body cast is applied and divided into 3 separate sections by making 2 encircling cuts in the plaster. The middle section is utilized to push laterally over the apex of the curve while the other 2 sections are pushed in the opposite direction. The correcting forces are continued while the cast is applied and hardens.

Surgical Treatment. It is necessary to determine with accuracy the extent of the fusion area and the degree of desired correction. The limits of the primary curve are ascertained because every vertebra in this arc must be fused Ideally, the end vertebrae should be parallel with one another. The primary curve should not be corrected to an angular value less than the angle of the compensatory curves after the latter's maximum capacity for spontaneous straightening has been obtained. Otherwise, the failure of the secondary curves to subside after spine fusion will result in bodily deformity in the opposite direction. The spontaneous correction in the compensatory curves is determined by the pelvic tilt film for the lumbar curve; the cervical and high curves in the dorsal area are studied by anteroposterior views after side-bending toward the side of convexity. The total values of the residual curves indicate the total deviation of the spine in this direction and represent the anticipated postoperative spontaneous correction. This value must not be less in the finally corrected and fused primary curve.

Preoperatively, an anatomic landmark is determined. A piece of metal is taped over a spinous process, and the site is also marked with gentian violet. An x-ray film reveals and identifies the vertebra. The number of vertebrae above and below this landmark to be included in the fusion is established.

A 24-hour sterile preparation is repeated in the operating room. An electric cast saw is held in readiness for emergency removal of the cast. Endotracheal anesthesia is used. Sufficient blood for replacement should be at hand. If the area to be fused is extensive, operation should be done in stages. No more than 6 to 8 vertebrae should be fused at one sitting, and the first procedure should be centered over the apex. The subsequent stages are at the extremities of the curve. When the deformity is extreme, it is technically difficult to expose the concave side. The technic consists essentially of the Hibb's procedure whereby multiple slivers of bone are elevated from the sides of the spinous processes and the back of the laminae and crossed over the interlaminar intervals. The facets are thoroughly curetted out. Cortical and cancellous bone removed from the ilium and the tibia are packed into both gutters but particularly on the concave side. This latter precaution utilizes the natural compression forces of the curve to stimulate bone formation and hypertrophy and to reduce the possibility of pseudarthrosis. If rib deformity is extreme, a rib may be removed and used as a graft.

Postoperatively, the patient is recumbent in the original cast for 3 months. Then a new well-fitted jacket is applied. The semibent jacket is preferred. Then ambulation is permitted, and the cast is reapplied at intervals over succeeding months until roentgenograms show a solid fusion. Nevertheless, studies and close observation should continue for evidence of pseudarthrosis and recurrence of the curve.

Pseudarthrosis is manifested by loss of correction, sense of fatigue, pain particularly radiating along an intercostal area, tenderness over the apex of the curve, a defect seen in the fusion mass and obvious change of position in bending films. If recurrence of the deformity is extreme, the turnbuckle cast should be reapplied, and the fusion site explored. Fibrous tissue is removed, the bony surfaces are freshened, and additional bone grafts for reinforcement are inserted. Postoperative treatment is the same.

Laminectomy is rarely done for decompres-

sion in severe scoliosis when paralysis threatens. Removal of a hemivertebra has been attempted in the lumbar region, but the operation is fraught with great danger, and the results are poor. The use of epiphyseodesis to retard longitudinal growth on the convex side is still in the experimental stage.

Scoliosis, even if well compensated, may be productive of painful degenerative arthritis in the adult. Spine fusion is indicated.

I

THE HOSPITAL FOR SPECIAL SURGERY

SCOLIOSIS HISTORY

Name	Date	History Number
Date of Birth	Age	Sex

Complaint: (High shoulder, prominent shoulder blade, high or prominent hip, prominent chest or breast, short leg, curvature, poor posture, awkward walk, pain in back, general fatigue, etc.)

Present History: Which deformity noted first

Age first noted — by whom (patient, parents, doctor, teacher, nurse)
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Duration of treatment
Progress of deformity (stationary, increasing, decreasing, rapid, slow)

Family History. Give history of scoliosis or other deformity in each case. (other side)

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				standing height		sitting height	
Father	Age			standing	height	sitting	height
Mother	Age			"	"	"	"
Brothers	1	Age		"	"	"	"
(names)	2	"		"	"	"	"
	3	"		"	"	"	"
	4	"		"	"	"	"
Sisters	1	"		"	"	"	"
(names)	2	"		"	"	"	"
	3	"		"	"	"	"
	4	"		"	"	"	"

Past History: Birth difficult or normal Evidence of trauma?

General health (robust, weak, normal weight, underweight, overweight, good color, pale, sickly, frequent illness) Illness—(age and duration) scarlet fever , measles , mumps chicken pox , whooping cough , polio , rickets, diphtheria, empyema , pneumonia , torticollis , Friedreich's ataxia chorea , tonsillitis menses, age onset, (regular, irregular, scanty, profuse, duration)

Examination Date

General posture (good, fair, poor) General development (good, fair, poor). Musculature (good, fair, poor) Nutrition (good, fair, poor). High shoulder (rt, left). Prominent scapula (rt, left). Prominent low ribs (rt, left) Exaggerated flank crease (rt., left). Prominent hip (rt, left). High hip (rt, left). List (rt., left). R. A. L. A Rotation (no, mild, moderate, severe)
Standing height Sitting height Weight
Flexibility of spine (flexible, moderately fixed, fixed)
Curve (Note muscle imbalance on other side of sheet)
Curve corrected on forward bending? (No, slight, moderate, complete)
Curve corrected on suspension? (No, slight, moderate, complete)

Diagnosis—Scoliosis (check which)

- A. Functional
- B Structural
  - I. Osteopathic
    - 1. Congenital
    - 2 Thoracogenic
    - 3. Other osteopathic
  - II. Neuropathic
    - 1 Congenital
    - 2 Post polio
    - 3. Other neuropathic
  - III. Myopathic
    - 1. Congenital
    - 2 Muscular dystrophy
    - 3 Other myopathic
  - IV. Idiopathic

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THE HOSPITAL FOR SPECIAL SURGERY

SCOLIOSIS RECORD

Date								
Age								
Ht Standing								
Ht Sitting								
L A								
R A								
List								
High Shoulder								
Low Hip								
Prominent Hip								
X-ray Date								
Born Age								
( ) Curve: Standing								
( ) Curve: Sitting								
( ) Curve: Supine								
Rotation (+, ++, etc)								
Wedging (+, ++, etc)								
( ) Curve: Standing								
( ) Curve: Sitting								
( ) Curve: Supine								
Rotation (+, ++, etc)								
Wedging (+, ++, etc)								
( ) Curve: Standing								
( ) Curve: Sitting								
( ) Curve: Supine								
Rotation (+, ++, etc)								
Wedging (+, ++, etc)								
( ) Curve: Standing								
( ) Curve: Sitting								
( ) Curve: Supine								
Rotation (+, ++, etc)								
Wedging (+, ++, etc)								

FIG 526 Scoliosis record. (Hospital For Special Surgery, New York City)

Special Situations. When congenital deformities of vertebrae and ribs are present, the spinal curve is quite fixed. Therefore, it is necessary to bend the secondary curve to balance the primary one and fuse it One must wait for cessation of growth.

Tilting of the 5th lumbar vertebra may be primarily due to some developmental anom-



Fig. 527 Congenital scoliosis The ribs on the concave side are misshapen and fused. The 7th thoracic vertebra is wedged

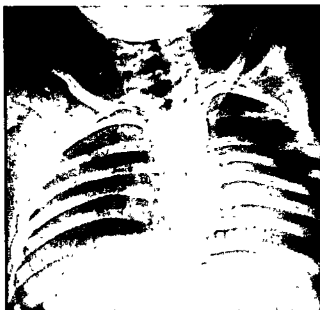


Fig. 528. Congenital scoliosis, showing wedged vertebrae, hemivertebrae and spina bifida occulta.

aly, and the lumbar curve as a consequence deviates to one side. The relationship of this vertebra to the pelvis remains unchanged regardless of bending, as demonstrated in pelvic tilt films Correction is obtained by bending the pelvis in the direction of accentuating the curve so as to align the superior border of the sacrum with the 3rd and the 4th lumbar and fusing If the tilt as shown on films is unstable but correctible, the 5th lumbar is first aligned with the superior sacral border, and lumbosacral fusion is done. In consequence, the secondary lumbar curve will straighten.

In 5th lumbar tilt secondary to a lumbar curve, the vertebra will straighten spontaneously with correction of the curve. However, if the lumbar curve is primary and relatively rigid, its correction can be accomplished better if the 5th lumbar is first aligned and fused to the sacrum and the wedging jacket used at a later procedure for the lumbar curve.[84]

CONGENITAL SCOLIOSIS; HEMIVERTEBRA; WEDGED VERTEBRA[85]

Three ossification centers, two in the vertebral arch and one in the body, appear at about the 8th week of fetal life in the cartilaginous anlage of the adult vertebra. Occasionally, the ossification center in the body may be bilobed or appear to be two distinct centers. The separate centers may remain separate throughout life as two distinct halves lying side by side, the so-called "hemivertebrae." Multiple symmetrically developed hemivertebrae are mechanically as efficient as normal vertebrae. However, when only one asymmetrically placed ossification center appears, one half of the body fails to develop, and a single hemivertebra results. The spine is unbalanced, and a scoliotic deformity re-

[84] Von Lackum, W H.: Surgical treatment of scoliosis *in* Bancroft and Marble: Surgical Treatment of the Motor-Skeletal System, p. 152, Philadelphia, Lippincott, 1951.

[85] Billig, E L.: Congenital scoliosis, an analytical study of its natural history, J. Bone & Joint Surg. 37A.404, 1955.

sults When a number of single hemivertebrae are balanced by an equal number of single hemivertebrae on the contralateral side, their effects neutralize each other, and the spine remains straight. Curvature develops only when a greater number of hemivertebrae are present on one side than on the other.

A wedged vertebra is defined as one in which the vertical height of the body on one side differs from that of the other side, as seen on the anteroposterior roentgenograms. It is due either to asymmetric ossification or to lateral compression secondary to scoliosis.

Most cases of "congenital" scoliosis are associated with congenital anomalies. Rarely can the deformity be classified as idiopathic. Single or multiple hemivertebrae are most often encountered. Wedged vertebrae may be found in conjunction with hemivertebrae; rarely are wedged vertebrae found alone with scoliotic deformity in early childhood. Anomalies of the ribs, particularly congenitally fused ribs, are frequently associated. These accentuate the curve to the opposite side. When a curve develops as a result of these anomalies, a compensatory curve usually forms, and the spine remains balanced. During the periods of rapid growth a rapid increase of primary and compensatory curves occurs, especially the upper and the mid-dorsal curves.

Treatment

Braces will not retard progression of deformity. During childhood, the spine is kept mobile by constant exercising. Changes in the degree of curvature are determined by x-ray study every few months. Because fused ribs almost invariably are associated with development of a severe curve, removal of the fused segments of ribs constitutes excellent prophylaxis. Particularly at the ages of puberty and thereafter, the spine must be watched closely. At the earliest sign of increase of the curve, jacket correction followed by spinal fusion is mandatory. Arthrodesis of the posterior spinal elements in a child will not interfere with longitudinal growth. Excision of a hemivertebra is not recommended.[86]

[86] Compere, E. L: Excision of hemivertebrae for correction of congenital scoliosis, J. Bone & Joint Surg 14.555, 1932.

28

The Pelvis

THE SACRO-ILIAC JOINT

The sacro-iliac articulation is formed by narrow, closely fitted, irregularly shaped and cartilage-covered surfaces of the postero-internal ilium and the lateral border of the sacrum. The joint is bridged by fibrocartilage (amphiarthrodial joint) or separated by a synovial cavity (diarthrodial joint) or consists of a combination of both types of joints. The investing ligaments include the anterior sacro-iliac, the posterior sacro-iliac and the interosseous sacro-iliac. The posterior sacro-iliac ligament is so thick and strong that violent trauma will produce a fracture to either side of the joint rather than a dislocation. The sacrotuberous ligament is a large, flat, triangular ligament which originates from the posterior inferior iliac spine and the posterior and the lateral surfaces of the sacrum and extends downward to attach to the tuberosity of the ischium. At the latter point, it becomes continuous with the tendon of origin of the long head of the biceps femoris. Thus, it becomes apparent that the hamstrings through their attachment to the tuberosity and acting via the sacrotuberous ligament exert a rotatory force which displaces the upper edge of the pelvis backward, straightens the lumbosacral angle and reduces lumbar lordosis.

Anteriorly, the lumbosacral trunk lies in direct relationship to the sacro-iliac joint. Therefore, an inflammatory neuritis is a not uncommon accompaniment of infectious arthritis The anterior ligaments are thin and easily distended by intra-articular swelling, easily palpated through the rectal wall

Posteriorly, the upper two thirds of the joint are covered by the posterior end of the ilium. The lower third of the joint is covered by the sacro-iliac ligaments but can often be palpated in thin individuals.

The conditions affecting the sacro-iliac joint are those which involve any joint. It is a favored site for tuberculous infection and often is the starting point for rheumatoid arthritis of the back. Degenerative arthritic changes are often pronounced at this joint Sacro-iliac strain and "subluxation" are controversial conditions, the existence of which is doubted by many.

Clinical Picture

Sacro-iliac joint involvement is typified by:

1. **Symptoms.** *Pain* is *local* over the joint or *referred,* usually to the groin and the posterior thigh. less often the leg. Pain is often increased by lying on the affected side.

2 **Findings.** The patient is most comfortable while sitting on the unaffected buttock. While standing, the tension of the hamstrings limits forward excursion of the pelvis; therefore, forward bending is limited and painful. While sitting, the hamstrings are relaxed, and forward bending is increased characteristically. The Lasegue sign causes pain over the joint. Gaenslen's test for sacro-iliac involvement is performed by hyperflexing the hip on the unaffected side, thereby firmly fixing that side of the pelvis The unaffected hip is then hyperextended. A torsion strain is exerted upon the sacro-iliac joint and is painful in the presence of disease. Externally, pressure over the lower third of the joint below the posterior inferior iliac spine may reveal tenderness Internally, on rectal digital examination, swelling and tenderness may be localized over the sacro-iliac joint. In the presence of an inflammatory condition, compression together of both iliac crests causes pain in many but not all cases. When inflammatory disease can be excluded, relief of pain by infiltrating the joint with a local anesthetic will define the location of the trouble accurately.

SACRO-ILIAC STRAIN

"Sacro-iliac strain" denotes painful stretching of the ligaments about the joint. The occurrence of this condition is regarded as uncommon because the sacro-iliac ligaments are very strong, and the movements of bending, lifting and hyperextension which produce a torsion strain upon the joint are more likely to cause a strain of thinner capsular ligaments surrounding the small lumbosacral joints However, there can be no question that sacro-iliac strain does occur and can be identified readily by an acute onset in the course of a torsion strain, tenderness over the joint, accentuation of pain by a maneuver which reproduces strain (Gaenslen's sign), and relief of symptoms by infiltration of the joint with a local anesthetic.

Certain circumstances favor strain of the sacro-iliac ligaments. The ligaments are softened and elongated by pregnancy, prolonged periods of bending and lifting and degenerative arthritis. The mechanism usually involves the act of straightening up from a stooped position It is tempting to suppose that muscular in-co-ordination is at fault. The hip flexors hold the ilium forward while the sacrum is rotated backward; or the hamstrings and the gluteus maximus extend the hip, rotating the ilium backward while the sacrum is held forward by the weight of the trunk. This theory seems to have support in the fact that a postural defect (pelvic inclination and excessive lumbar lordosis) is an associated finding.

"Sacro-iliac subluxation" implies that ligamentous stretching has been sufficient to permit the ilium to slip on the sacrum. An irregular prominence of one articular surface becomes wedged upon another prominence of the apposed articular surface, the ligaments are taut, reflex muscle spasm is intense, and pain is severe and continuous until reduction is effected. The displacement is so slight that it cannot be recognized in roentgenograms. The condition is differentiated from sacro-iliac strain in that pain is more intense and not relieved by sitting, recumbency, or a tight encircling bandage. The pain of subluxation is often relieved dramatically and suddenly by manipulation.

Treatment

Sacro-iliac strain responds to bed rest, heat and a firmly constricting elastic bandage which encircles the pelvis just below the level of the iliac crests. Repeated injections of a local anesthetic about the joint posteriorly seems to expedite recovery. Avoidance of occupations requiring excessive bending and lifting is particularly necessary when degenerative changes are present.

Sacro-iliac subluxation may be reduced by manipulation. Either local or general anesthesia may be used. The maneuver is performed as follows: The patient lies on the unaffected side. An assistant forces the shoulder on the affected side forward toward the table. At the same time, the operator exerts a rotatory force by pushing the ilium in the opposite direction. The hip should be maintained in flexion during the procedure. Very often a palpable and audible snapping sound is perceived which indicates reduction. When the maneuver is successful, severe pain is relieved immediately, and substantial reduction in degree of response to straight-leg raising is observed. Subsequently, only the soreness and the tenderness of damaged ligaments remains. A constricting bandage should be worn for 3 weeks to reduce the possibility of recurrence.

Repeated incidents indicate undue laxity of ligaments. Injection of a sclerosing agent (Sylnasol) about the posterior aspect of the joint may occasionally immobilize the joint effectively. The injection of a proliferative substance is often followed by intense pain which must be controlled by narcotics and icepacks.

With excessive mobility and frequent joint displacements, the articular surfaces are eroded, and a traumatic degenerative arthritis ensues. Arthrodesis is necessary to eradicate symptoms permanently. It is advisable to fuse both sacro-iliac joints and the lumbosacral junction (trisacral fusion).

COCCYGODYNIA

The coccyx is the rudimentary tailbone of lower animal forms. It is a cone-shaped section of the spine composed of 4, occasionally

5, segments. These segments are separate and movable at birth but tend to fuse—the distal ones in childhood, the proximal ones in early adulthood. The sacrococcygeal junction remains movable throughout life but rarely may fuse. The coccyx is bound to the lower end of the sacrum by the sacrococcygeal ligaments, which attach to the margins of the sacral hiatus. Along the lateral borders of the coccyx the coccygeal muscles are attached; they extend laterally and forward to insert on the ischial spine. A strong fibrous ligament, the anococcygeal ligament, attaches to the tip of the coccyx and extends forward to attach to the external sphincter of the anus. The levator ani muscles lie in front of the coccyx and descend to insert into the anococcygeal ligament and the internal sphincter. One of the actions of the coccygeal and the levator ani muscles is to hold the coccyx forward. The nerve supply of the coccyx arises from the posterior primary divisions of the coccygeal nerves which emerge from the sacral hiatus. Each nerve receives a communicating branch from the posterior division of the lowermost sacral nerve. The main blood supply of the coccyx is derived from the middle and the 2 lateral sacral arteries which descend on the anterior aspect of the sacrum close to the bone. The coccygeal body, the glomus coccygeum or gland of Luschka, lies in front of the tip of the coccyx.

Because of its muscular attachments, the coccyx is in constant motion, particularly in the act of defecation. Pressure is exerted against the posterior aspect of the bone in sitting, the coccyx acting as a shock absorber and moving forward.

Clinical Picture

Pain about the coccyx results from local conditions or is referred from other regions.

1. Local Causes

A. *Sprain of Sacrococcygeal Ligaments.* A kick, a fall on the buttocks, or obstetric trauma will strain or tear these ligaments. The condition becomes chronic because the acts of sitting and defecation continually strain the already injured ligaments. A constant annoying discomfort is experienced and accentuated when sitting on a hard surface or during defecation. Occasionally, bending forward is painful. The sacrococcygeal joint is tender, and movement of the coccyx on rectal examination reproduces the pain. Injection of a local anesthetic about the joint affords immediate relief, and repeated injections may effect a cure. Hot sitz baths are prescribed, and a rubber ring or pillow is used when sitting. Manipulation of the coccyx under anesthesia by moving the joint through an extreme range of motion stretches the ligaments so that ordinary movements will no longer be painful. Resistant cases are cured by coccygectomy. The lower end of the sacrum must be removed to avoid a painful pressure area. Chronic sprain of the sacrococcygeal joint with repeated subluxation may eventuate in a localized traumatic degenerative arthritis.

B. *Degenerative Arthritis of the Sacrococcygeal Joint.* A forceful blow or severe obstetric trauma may cause subluxation of the sacrococcygeal joint. Gradually over several years narrowing, irregularity and sclerosis develop. Movement of the joint is restricted, and passive movement of the coccyx reproduces pain at the sacrococcygeal joint. When the individual sits on a hard surface, the coccyx fails to give, and a painful pressure point develops over the tip of the bone. Needling of the tip with a local anesthetic relieves the discomfort. If pain is localized to the sacrococcygeal joint, removal of the coccyx and the lower end of the sacrum is necessary.

C. *Fracture and Dislocation.* The effect of the attached musculature is to pull the distal segments forward. No subsequent symptoms are associated unless a painful pseudarthrosis or a degenerative arthritis with a stiff sacrococcygeal joint develops. This necessitates coccygectomy.

D. *Pain at the Tip of the Coccyx.* Occasionally, even in the absence of a rigid coccyx, a painful tender nodule is found at the distal end of the coccyx at the site of attachment of the anococcygeal ligament. Repeated procaine needling causes disappearance of the nodule and effects a cure.

E. *Tumors.* The frequency of a chordoma, a destructive tumor, in the lower end of the sacrum and the coccyx should be borne in mind.



Fig 529. Dislocated coccyx. A cause of persistent coccygodynia. Cure effected by coccygectomy.

2. Referred Causes. Injection of a local anesthetic about the coccyx fails to relieve the pain when referred from other regions.

A. *Lumbosacral Lesions.* An unstable 5th lumbar lamina of spondylolisthesis will compress the dura and its contained nerve roots and cause coccygeal pain. A large protruded mid-line disk is said to cause coccygodynia similarly Immobilization of the lumbosacral spine may relieve the pain and define the causative lesion.

B. *Episacral Lipoma.*[1, 2, 3] Fibrofatty nodules, because of edematous swelling or herniation through the deep fascia, can cause referred pain. Coccygodynia is associated with lipomata situated over the posterior aspect of the sacrum Injection of a local anesthetic relieves the pain about the coccyx Repeated puncturing or surgical removal of the lipomata effects a cure.

[1] Dittrich, R J Coccygodynia as referred pain, J Bone & Joint Surg 33A 715, 1951.

[2] Copeman, W C Fibro-fatty tissue and its relation to rheumatic syndromes, Brit M J 2 191, 1949

[3] Copeman, W S. C, and Ackerman, W L. Edema or herniations of fat lobules as a cause of lumbar and gluteal "fibrositis," Arch Int Med. 79 22, 1947.

OSTEITIS PUBIS[4, 5]

Osteitis pubis is a painful condition about the symphysis pubis, usually developing after surgical trauma in adjacent areas, characterized by bony resorption about the symphysis and spontaneous reossification with subsidence of symptoms.

Etiology

Symptoms develop within a few days or weeks after trauma, usually surgical, about the lower pelvic region. In males, a suprapubic or retropubic prostatectomy most commonly antedates symptoms. In females, pain often develops during the postpartum period, but the onset may occur during pregnancy or after extensive pelvic surgery.

The actual cause is unknown. Infection has been blamed, but local and general inflammatory signs are absent. Softening of the pelvic ligaments such as occurs during pregnancy suggests a predisposing factor.

Clinical Picture

Within a period of a few days, an excruciating pain develops about the symphysis pubis and the pubic rami and radiates along the adductor aspect of both thighs. Pain is intensified by any movement of the extremities, particularly abduction, which puts the abductor muscles on a stretch. The sites of attachment of the adductors are particularly tender, although the bodies and the rami of both pubes are diffusely tender. The area lacks the swelling, the redness and the warmth of inflammation, and the temperature is normal. Injection of a local anesthetic into the points of attachment of the adductors frequently relieves the pain.

The patient is thoroughly disabled and bedridden. Often he seeks a position of maximum comfort, which is one of flexion and adduction. The pain is intense for a period varying from days to weeks and then gradually subsides over an indefinite period from a few months to several years.

[4] Beer, E (Osteitis pubis) periostitis of the symphysis and descending rami of the pubis following suprapubic operations, Int J. Med Surg 37:224, 1924

[5] Wiltse, L L, and Frantz, C. H.: Non-suppurative osteitis pubis in the female, J. Bone & Joint Surg 38A 500, 1956.

The author has seen many cases with typical symptoms, but lacking in x-ray findings, which rapidly improved within a few weeks. These may constitute mild abortive types.

ROENTGENOLOGIC FINDINGS

Within the early days or weeks, roentgenograms are negative. As the disease progresses, the bone adjacent to the symphysis undergoes spotty demineralization which intensifies until the symphysial gap appears to be widened. Characteristically, the opposing aspects of the pubic bodies are moth-eaten, rarefied and cup-shaped. The rami are diffusely osteoporotic. Gradually, over many months, reossification occurs with restoration of bony architecture. Occasionally, the symphysial interval is narrowed or completely obliterated.

PATHOLOGY

Reports in the literature vary. Wiltse and Frantz demonstrated degenerative changes in the fibrocartilage of the symphysis. The condition may in some way be related to partial separation of the sites of attachment of the adductor tendons. This is suggested by the complete relief obtained by local anesthetization of these areas.

TREATMENT

The condition shows a tendency to spontaneous cure. Therefore, various forms of therapy, notably irradiation, are of doubtful efficacy. The attack seems to be shortened by *injecting a local anesthetic subperiosteally* into the tender sites and traumatizing the osseotendinous junctures by multiple needle punctures. The patient is placed at absolute bed rest, the lower extremities are placed close together and flexed over a pillow. Maximum relief of pain is obtained by icebags, but narcotics are often necessary.

OSTEITIS CONDENSANS ILII[6, 7]

Osteitis condensans ilii is a disturbance of the normal architecture of the ilium in which increased condensations of bone occur in the auricular portion of the ilium without a corresponding change in the sacro-iliac joint or the sacrum.

[6] Hare, H F., and Haggart, G E. Osteitis condensans ilu, J.A.M.A 128·723, 1945.

[7] Shipp, F L, and Haggart, G. E.: Further experience in the management of osteitis condensans ilii, J. Bone & Joint Surg. 32A·841, 1950.

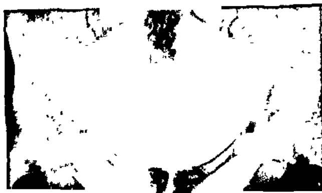


FIG. 530. Osteitis pubis. Osteolytic areas bordering on the symphysis are well displayed. Surrounding increased density of the body of the pubis represents reactive bone formation replacing the osteolytic lesions. (Dr. W. J. Schnute's case)

ETIOLOGY

The actual cause is unknown. Theories include:

1. **Mechanical Strain.** The sacrum tends to rotate about a fulcrum situated about the 2nd sacral segment. The strong sacro-iliac ligaments become taut and resist this rotation. When tendency to rotation is increased because of an *acute lumbosacral angle* or during *pregnancy* when the pelvic ligaments soften, additional strain is thrown upon the *ligaments at their attachment to the ilium.* The auricular process of the ilium responds by bony thickening.

2. **Aseptic Necrosis.** This seems to be unlikely in view of the bilateral involvement, and the microscopic appearance of living bone.

PATHOLOGY

Microscopically, the trabeculations are greatly increased in thickness. Focal areas of fibrosis occur in the marrow. Osteoblastic activity proceeds normally or in excess. Osteocytes are present within their lacunae. Scattered islands of cartilage are observed at a considerable distance from the joint surface.

SYMPTOMS

Chronic low back pain is persistent, never severe, and radiates to one or both buttocks,

never into the sciatic distribution. Symptoms are *aggravated by activity* and *relieved by rest*. There is no night pain. The condition occurs in females of childbearing age, frequently having its onset in the final trimester of pregnancy or immediately after delivery. Symptoms recur or are exaggerated during subsequent pregnancies.

Physical Findings

Lumbar lordosis is increased. Spasm of the erector spinae muscles may be present. Tests for sacro-iliac disease are negative. Musculature is inadequate and frequently is associated with obesity. The erythrocyte sedimentation rate is within normal limits.

Roentgenologic Findings

A flat anteroposterior sacral projection, preferably stereoscopic, reveals condensation of the auricular portion of the ilium. This area lies behind the anterior margin of the sacro-iliac joint. The roentgenographic picture is not proportionate to the severity of symptoms

Differential Diagnosis

Marie-Strümpell arthritis causes bone condensations about the sacro-iliac joint; however, they occur on both sides of the joint. The male is involved more commonly. Sacro-iliac tests are positive. The ESR is elevated. Roentgenograms show roughening, apparent widening, destruction, and eventual obliteration of the joint by bony fusion. Osteitis condensans ilii occurs only in the female and only in the ilium; the joint is free of involvement; and the ESR is normal.

Treatment

Attempts are directed to reducing the lumbosacral angulation by posture exercises Weight reduction, a bedboard or a hard mattress, and a lumbosacral corset are prescribed Symptoms are usually eliminated by this regimen, and occasionally the sclerosis may actually diminish. Rarely, surgical arthrodesis may be indicated.

Addenda

Many deny the existence of this entity. Rather, they believe that the condensations are a coincidental finding in cases of lumbosacral strain.

EPISACRO-ILIAC LIPOMAS[8]

Small nodules of fibrofatty tissue about the posterior iliac crest and over the sacrum are *occasionally the cause of low back pain*. Normally, a layer of fibrous adipose tissue lies in this region, covered by a layer of deep fascia. When a defect develops in the fascia, the fibrofatty tissue herniates through the small opening, becomes edematous and causes pain. Just above the iliac crest alongside the sacrospinalis are several foramina in the fascia through which the posterior rami of the last 3 lumbar nerves and small blood vessels reach the integument. Fatty nodules often protrude through these openings.

Clinical Picture

A vague pain develops gradually about the iliac crest and the buttock and is aggravated by stooping and by direct pressure over the lower back. The nodules are palpable as soft movable tumors which slip beneath the examining finger. Pressure over the nodule reproduces the discomfort and often radiation to the posterior aspect of the thigh. Nodules over the mid-line of the sacrum cause pain *referred toward the coccyx*. Infiltration of the fatty tumor with a local anesthetic relieves the symptoms.

Treatment

Under local anesthesia the fatty nodule is destroyed by multiple punctures with a large-bore needle. To ensure its disintegration, the area should be forcibly massaged. Surgical removal is rarely indicated.

TUBERCULOSIS OF THE SACRO-ILIAC JOINT[9, 10]

Tuberculous infection of the sacro-iliac joint frequently is associated with tuberculo-

[8] Copeman, W. S. C, and Ackerman, W. L Edema or herniations of fat lobules as cause of lumbar and gluteal "fibrositis," Arch Int Med 79.22, 1947

[9] Pollack, N, and Bosworth, D M. Sacro-iliac tuberculosis, Quart. Bull Sea View Hosp 7.415, 1942

[10] Soholt, S. T: Tuberculosis of the sacro-iliac joint, J. Bone & Joint Surg 33A 119, 1951

sis of the spine at the lumbosacral area and of the hip, suggesting the case of spread from one to the other, perhaps via the psoas muscle. Destructive caseous osseous lesions are the rule and often destroy the joint and form abscesses. The abscess may present dorsally over the joint or intrapelvically, erupting at the inguinal region. Rupture of the abscess results in a resistant sinus and secondary infection. Severe visceral lesions are almost invariably associated, resulting in a serious outlook. If the patient survives, spontaneous bony ankylosis of the sacro-iliac joint almost always occurs after 3 or 4 years. The disease may be bilateral.

Clinical Picture

Age. Young adults. Rare in infancy and childhood

Onset. Gradual; may follow trauma or pregnancy.

Symptoms. Pain over the joint, referred most often to the groin, less commonly along the sciatic distribution. The pain is accentuated by direct pressure as by recumbency, particularly when turning about in bed. Sitting on the buttock on the affected side is painful, whereas sitting on the opposite buttock relieves the pain. Bending forward with the knee extended is painful, whereas bending with the knees flexed is painless Jarring as in walking, coughing, sneezing accentuates the discomfort

Findings. The patient lists to the opposite side. With the lower extremities extended, forward bending is limited. When the knees and the hips are flexed, the hamstrings are relaxed, tension is removed from the pelvis, and further forward bending is accomplished. Only the lower end of the joint is superficial posteriorly and displays tenderness and perhaps a boggy swelling The swelling and the tenderness may be more demonstrable on rectal examination Compressing the iliac crests together causes direct painful pressure on the joint *Genslen's sign* for sacro-iliac disease depends upon twisting the ilium on the sacrum. It is performed by first flexing the hip strongly on the unaffected side. This firmly fixes the pelvis and the lumbosacral junction The opposite hip is then hyperextended, thereby rotating the corresponding ilium backward. The strain on the inflamed ligamentous structures about the sacro-iliac joint supposedly produces the pain. The low-grade inflammatory swelling of a cold abscess or a sinus may be present.

Roentgenologic Findings

Early changes consist of haziness or loss of definition of the joint line. Next, an irregularity of the articular surface with areas of erosion develops. If the disease subsides, the joint is narrowed, but the adjacent bone is dense. If destruction progresses, marked cavitation is evident. Later changes show a bony ankylosis developing in the vast majority of cases. The remainder of nonfused cases show only increased density of the articular bone.

Prognosis

Before antibiotic therapy, sacro-iliac tuberculosis was synonymous with generalized severe infection, and mortality was high. A draining sinus never healed unless the joint fused. Now this disease constitutes only one of many foci which can be eradicated surgically to improve the outlook greatly. Although bony ankylosis is almost inevitable after several years in patients who survive, it is necessary to eliminate this disease as quickly as possible. Rarely is conservative treatment indicated and only when the infection remains localized to this region.

Treatment

Conservative treatment consists of prolonged immobilization in a cast which extends from the nipples to the knees. If a sinus is present, immobilization on a bilateral Thomas abduction frame is adequate. Streptomycin, PAS and INAH are given in addition to general measures. Bony obliteration of the joint indicates a cure and may require 1½ to 2 years

Surgical treatment is aimed at arthrodesis of the sacro-iliac, either intra-articular or extra-articular.

Intra-articular Arthrodesis (Smith-Petersen). An incision is made along the posterior two thirds of the iliac crest then extended outward and downward parallel with

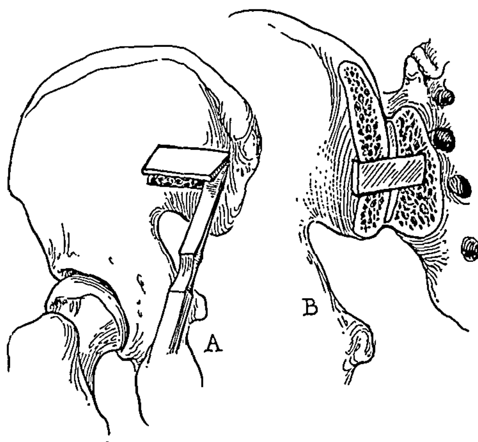


Fig. 531. Arthrodesis of the sacro-iliac joint. (Smith-Petersen)

the fibers of the gluteus maximus. The muscle is elevated subperiosteally, exposing the lateral aspect of the ilium. A rectangular block of bone is removed from the ilium directly over the sacro-iliac joint To locate the joint, a line is drawn from the anterosuperior spine to the posterosuperior spine. At a point 1½ inches lateral to the posterior superior spine a perpendicular is dropped for ½ inch and is the site of the joint. Upon removal of the block of bone the articular surface of the sacrum is exposed The articular cartilage and all abnormal tissue is removed from the block and the sacrum until cancellous bone presents. The block is replaced and countersunk until firm approximation of cancellous surfaces is obtained. Several slivers of bone may be turned over from the ilium over the block to promote osteogenesis. The muscle is sutured back into position.

Extra-articular Arthrodesis. An incision is made along the posterior half of the iliac crest to the posteroinferior spine. The soft tissues are elevated from the outer and the inner aspects of the ilium. The erector spinae is elevated from the sacrum and is retracted medially. A large single block of bone is removed from the exposed iliac crest The sacrum and the adjacent inner aspect of the ilium are denuded until cancellous surfaces are exposed. Into this angle are placed the large bone graft and multiple small cortical and cancellous chips removed from the lateral iliac surface. The gluteus muscle sutured back to the erector spinae holds the bone grafts snugly in place.

PART FOUR

Special Subjects



29

Radioactive Isotopes in Orthopaedics

RENATO BASERGA, M.D.*

The use of radioactive isotopes in the investigation and the treatment of orthopaedic conditions, although in its early stages, promises unlimited possibilities of development. The following material is offered as a general basis for study by the orthopaedic resident and surgeon, with a hope that it will elicit some interest in this fascinating new field.

To physicians, radioactive isotopes are important in 3 ways: (1) as agents of disease; (2) as tracers in biochemical investigations, especially metabolic studies; and (3) as tools in the diagnosis and the treatment of illnesses. These 3 facets will be considered separately, after a brief review of the nature of isotopes and their general properties.

THE NATURE OF ISOTOPES

ATOMIC STRUCTURE

According to modern physics, atoms are thought of as complex aggregations of positive and negative electrical units.[1] There are several types of subatomic particles, but a sufficiently clear understanding of atomic structure can be obtained by considering atoms as composed of (1) the electron, which has a negligible mass and carries a unit negative electrical charge; (2) the proton, which has a unit mass and carries a unit positive electrical charge, and (3) the neutron, which has the same mass of a proton but no electrical charge Each atom consists of a nucleus, made up of protons and neutrons and surrounded by free electrons arranged in concentric orbits. In the whole atom there is always an equal number of protons (positive charges) and electrons (negative charges), so that the atom itself is electrically neutral or uncharged. Figure 532 shows the structure of the atoms of hydrogen, boron, carbon and oxygen. It will be noted that the number of protons in the nucleus always equals the number of electrons in the various orbits.

Each atom is distinguished by an atomic number and an atomic weight. The atomic number, which characterizes the chemical properties of an element, is determined by the number of positive charges carried by the nucleus (or by the number of planetary electrons, which is the same). For example, in Figure 532, hydrogen, with only one proton, has an atomic number of 1; boron, with 5 protons, has an atomic number of 5, etc.

The atomic weight is the weight of an atom, referred to the atom of oxygen taken as a standard. The weight, or mass, of an atom is due chiefly to its protons and neutrons, because electrons have a negligible mass. The atom of oxygen, with 8 protons and 8 neutrons, is said to have a weight of 16. From Figure 532, it can be seen that hydrogen, with only one proton, will have an atomic weight of 1; boron, with 5 protons and 6 neutrons, will have an atomic weight of 11; and carbon, with 6 protons and 6 neutrons, will have an atomic weight of 12. In general, as the atomic number is given by the number of protons, and the atomic weight by the number of protons and neutrons, it follows that the number of neutrons is equal to the atomic weight less the atomic number.

ISOTOPES

It is now known that atoms of the same element, that is with the same number of

* Department of Pathology, St Luke's Hospital, Chicago, Ill

[1] Glasstone, S Sourcebook on Atomic Energy, Princeton, N J, D Van Nostrand Co, Inc., 1958

1p

ATOMIC NUMBER 1
ATOMIC WEIGHT 1

5p
6n

ATOMIC NUMBER 5
ATOMIC WEIGHT 11

6p
6n

ATOMIC NUMBER 6
ATOMIC WEIGHT 12

8p
8n

ATOMIC NUMBER 8
ATOMIC WEIGHT 16

FIG. 532. Structure of the atoms of hydrogen, boron, carbon and oxygen. In the nucleus, p stands for proton and n for neutron; electrons are represented by black dots outside the nucleus. Note that the atomic number is that of the protons in the nucleus, while the atomic weight is the sum of both protons and neutrons.

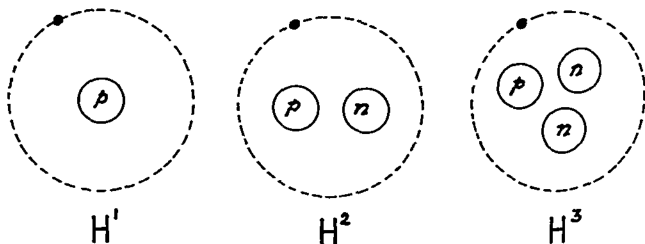
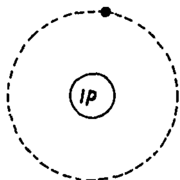
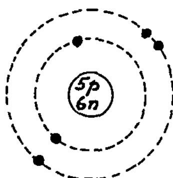


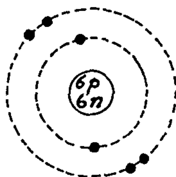
FIG. 533. Isotopes of hydrogen. The number of protons p and electrons o is always the same, but the number of neutrons n varies from 0 to 2.

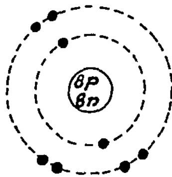




ATOMIC NUMBER 1
ATOMIC WEIGHT 1

ATOMIC NUMBER 5
ATOMIC WEIGHT 11

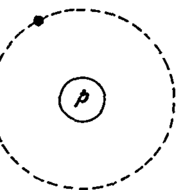


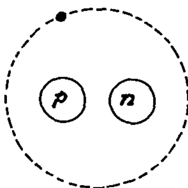


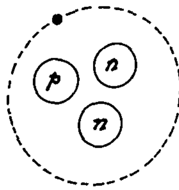
ATOMIC NUMBER 6
ATOMIC WEIGHT 12

ATOMIC NUMBER 8
ATOMIC WEIGHT 16

FIG 532 Structure of the atoms of hydrogen, boron, carbon and oxygen. In the nucleus, *p* stands for proton and *n* for neutron; electrons are represented by black dots outside the nucleus. Note that the atomic number is that of the protons in the nucleus, while the atomic weight is the sum of both protons and neutrons.







H^1 H^2 H^3

FIG. 533. Isotopes of hydrogen. The number of protons *p* and electrons *o* is always the same, but the number of neutrons *n* varies from 0 to 2.

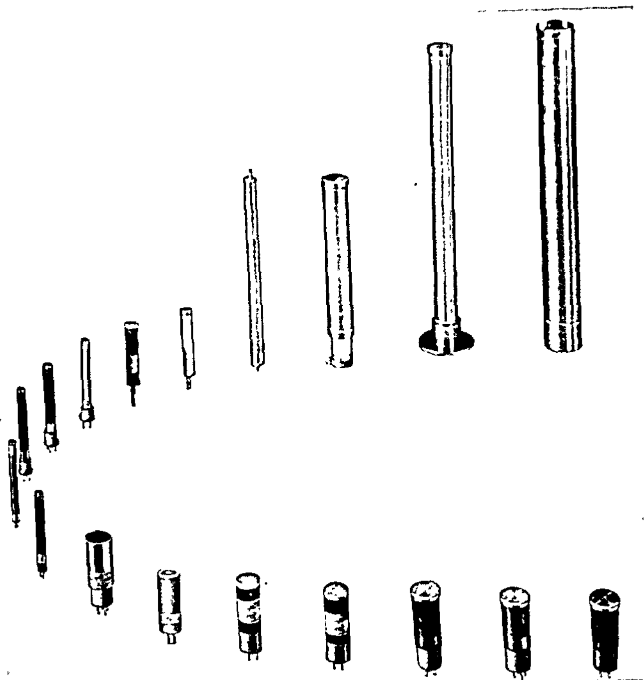


FIG. 535. Detectors are the modern version of the original Geiger-Mueller tubes They come in many sizes and shapes for various purposes. (Tracerlab Inc.)

Thus, their detection is simply a problem of detecting beta and gamma radiations. For this purpose the best available instruments are the discharge counters,[6,7] which are the modern version of the original instrument developed by Geiger and Mueller. The discharge counters come in a great variety of sizes and shapes, and the choice of a particular counter depends upon the desired application, since there is no universal Geiger-Mueller tube which is optimum for all applications (Fig. 534). The theory upon which the Geiger-Mueller counters are built is complicated: briefly, they consist of a glass chamber, containing a diode and filled with gas at a low pressure. The beta and the gamma rays enter the counter through a mica window, interact with the gas

[6] Rovner, L Geiger counters *in* Glasser's Medical Physics, vol 1, p 486, Chicago, Year Book Pub, 1944

[7] Glasstone, S. Sourcebook on Atomic Energy, Princeton, N. J., D. Van Nostrand Co, Inc, 1958



FIG. 534. Autoradiograph of femur of rabbit injected with Strontium^{90}. Note periosteal deposition of Strontium^{90} (blackened area) and pathologic bone formation above Rabbit 7 weeks old injected with 1,000 microcuries per kilogram of Strontium^{90}. Died 6 months later. (From Dr. Jenifer Jowsey, Argonne National Laboratory, Lemont, Ill.)

far from unity, result in stable nuclei A large number of radioactive isotopes can now be prepared in the nuclear reactor by the above-described process or by similar ones [3]

PROPERTIES OF RADIOACTIVE ISOTOPES

Radioactive isotopes must be regarded as chemically and biologically identical with the stable isotopes of the same substance.[4] Each radioactive isotope is characterized by: (1) the type and the energy of radiation emitted. and (2) its lifetime or rate of decay.

As mentioned above, radioactive isotopes emit 3 kinds of radiation, known as alpha, beta and gamma rays. Alpha rays are streams of double-charged helium nuclei He^4 (atomic number: 2, mass: 4). They are emitted by many heavy radioactive elements such as radium, uranium and plutonium, but not by the tracer elements that have recently become important in biology and medicine. Beta rays can be either positive or negative. The latter are high-speed electrons with negative charge and are emitted by a large variety of radioactive elements. Positive beta rays are positively charged electrons (or positrons) and have only a transitory existence. Gamma rays are electromagnetic radiations, a term which includes radio waves, infrared rays, visible light, ultraviolet rays and roentgen rays and may be considered as x-rays of short wave length. Their emission usually follows the emission of alpha or beta particles, leaving the nucleus of the atom in a state of excitement. The transition from this "excited" energy state to a lower energy state, called isomeric transition, is responsible for the emission of gamma rays. Because of their common mode of action, alpha, beta, gamma and x-rays are known as ionizing radiations. The effects of ionizing radiations on living organisms will be considered later.

Radioactive atoms, by emitting particles, spontaneously decay until they become stable. Their rate of decay is measured by the so-called half-period or half-life of the element. Whereas the total life of a radioactive element is a useless number, as it may approximate infinity, the half-life is a very useful characteristic. Each radioactive element has its own characteristic half-life, which is defined as the time required for one half of the atoms to decay. The half-lives of various radioactive atoms vary from fractions of a second to millennia. For example, Silver-110 has a half-life of 24.2 seconds, while Carbon-14 has a half-life of 5,568 years. The half-life is of great importance in determining the medical usefulness of a radioisotope.[5] Those with short half-lives are uneconomical to ship long distances, and those with long half-lives are potentially carcinogenic. The most suitable in medicine, with some exception, should have a half-life range between 10 hours and 30 days

DETECTION AND MEASUREMENT OF RADIOACTIVE ISOTOPES

As mentioned above, radioactive isotopes are chemically indistinguishable from their stable counterparts. However, they emit radiations that can be detected and measured quantitatively. Nearly all artificial radioactive isotopes used in medicine and in biology emit beta rays, and some also emit gamma rays.

[3] Radioisotopes, Special Materials and Services, Oak Ridge, Tenn., Oak Ridge National Laboratory.

[4] Evans, R. D : *in* Glasser's Medical Physics, vol 1, p. 643, Chicago, Year Book Pub., 1944

[5] Rosenthal, D J., and Lawrence, J. H.: Radioactive isotopes in medicine, Ann Rev. Med. 8:361-388, 1957.

radioactivity of a given material is thus reported in counts per minute per milligram, or counts per minute per millimole, and is called specific activity of that material. This measurement is the one most frequently encountered in medicine and is a relative measurement, which will perhaps amuse the physicist but is accurate enough for most biologic investigations.

Radioactive isotopes are also detected qualitatively by autoradiography, in which a photographic emulsion is used to obtain a visible representation of the distribution of radioisotopes in a variety of subjects.[8, 9] In medicine, autoradiography is used mainly to localize radioactive isotopes in tissue sections: a strip of photographic film is applied to a tissue section for a variable time and then developed. Afterward the section is stained by the usual methods, and by superimposing the photographic strip on the stained section, it is possible to localize the deposits of radioactive material (Figs. 535-538).

[8] Norris, W. P., and Woodruff, L. A.: The fundamentals of radioautography, Ann. Rev. Nuclear Sc. 5:297-326, 1955.

[9] Gross, J., Bogoroch, R., Nadler, N. J., and Leblond, C. P.: Theory and methods of the radioautographic localization of radioelements in tissue, Am. J. Roentgenol. 65:420-458, 1951.

RADIOACTIVE ISOTOPES AS AGENTS OF DISEASE

The physical and the chemical properties of the radioactive and the stable isotopes of all but the lighter elements are virtually indistinguishable, except for the radiation emitted by the radioisotope. No physiologic differences between the radioactive and the stable isotopes are evident, so long as the intensity of the radiation is kept below a level that

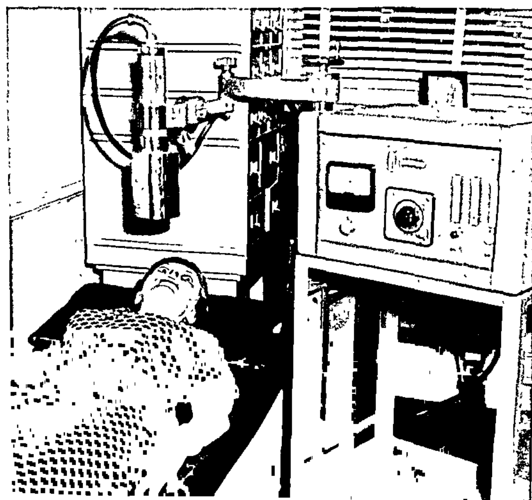


Fig. 538. A discharge counter in clinical operation. The detector above the head of the patient is supported by and connected to a scaler. (Tracerlab Inc.)

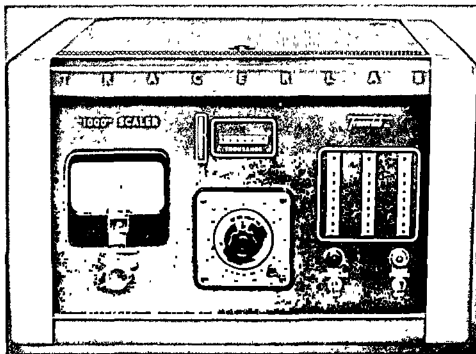


FIG. 536. Radiations detected by the Geiger-Mueller tubes are amplified, registered and tallied by a scaler, which can usually serve several types of detectors. (Tracerlab Inc.)

contained therein and form ions, which cause a discharge of electrical current between a highly positive central wire and a cathode plate. The electrical current produced across the interelectrode space is amplified and registered in a counting device and tallied to give the total number of discharges. As the number of electrical discharges is proportional to the concentration of radioactive atoms, a quantitative measurement can be obtained. The



FIG. 537. A discharge counter in operation in a laboratory. The detector, held in place by supporting equipment, is connected to the scaler. (Tracerlab Inc.)

with severe damage of the bone marrow. In more chronic cases, the skeletal lesions appeared first as discrete or confluent areas of bone destruction and rarefaction, which were slowly progressive and were complicated in several instances by malignancies developing in the damaged skeleton. Similar lesions have been observed in a number of patients who, in the 2nd and the 3rd decades of this century, were treated with radium salts for a great variety of illnesses. Thus, radium poisoning although a rare disease, is now a well-recognized clinical and pathologic entity, which the physician should always consider in the investigation of a disease In this connection, it is of special interest to know that heavy elements, such as uranium, plutonium and other transuranic elements of the actinide series, as well as radioactive isotopes which are formed in abundance in the fission of uranium or plutonium, are deposited and fixed in the skeleton. Though bone sarcomas have not been reported yet in man, tumor formation in bone following the administration of plutonium and of various fission products, notably strontium (Sr^{89}), has been reported in experimental animals and has suggested the possibility that these bone-seeking isotopes may produce similar harmful effects in man.[17]

RADIOACTIVE ISOTOPES AS TRACERS

A tremendous volume of literature has accumulated in the past few years over the use of radioactive isotopes as tracers in biochemical research.[18, 19] Briefly, in this type of investigation, one takes advantage of the radioactivity of a compound to follow its distribution, concentration and fate in the animal body. Most of the artificial radioactive isotopes can be used for such purpose, but the ones most frequently employed are Deuterium (H^2), Carbon^{14}, Nitrogen^{15}, Na^{24}, P^{32}, S^{34} and S^{35}, Fe^{55}, Co^{60}, Cu^{64}, Zn^{65} and I^{131}. The types of biochemical problems that are best studied by the use of tracers are essentially three: (1) anatomic distribution of a compound; (2) precursor-product relationships; and (3) analysis of rates of processes.

By administering isotopic material to the animal body, it is possible, by subsequent analysis, to ascertain its distribution in the various tissues or products and, by analogy, the distribution of its stable counterpart. The localization of the isotopic atom can even be established at the cellular level by the use of the technic of autoradiography, mentioned above. This type of investigation has been particularly useful in determining the distribution in the body of administered compounds and/or their products, such as water, electrolytes,[20] carcinogenic hydrocarbons,[21] and others.

The purpose of experiments dealing with the precursor-product relationship is to determine whether compound A can be converted into B in the organism. This can be accomplished by analyzing for isotope the carefully purified compound B after administration of isotopic compound A. This technic has given dramatic results in the investigation of the biosynthesis of various components of the animal body, such as steroids, amino acids, nucleic acids and hemoglobin.[22] Conversely, the same technic applies to the study of the catabolism of these compounds and/or others, such as carbohydrates and fatty acids.

Finally, from the use of isotopes in the analysis of rates of processes has evolved the modern concept of continuous turnover (synthesis, degradation and replacement) of certain body constituents (proteins, depot lipids, bone minerals, etc.) hitherto considered to be relatively stable in their composition.

These different technics are naturally applicable to the investigation of bone metab-

[17] Lisco, H.: Bone as a critical organ for the deposition of radioactive materials *in* Ciba Foundation Symposium on Bone Structure and Metabolism, pp 272-282, London, Churchill, 1956

[18] U S Atomic Energy Commission Isotopes. An Eight Year Summary of United States Distribution and Utilization, Government Printing Office, Washington, D C, 1955

[19] Ciba Foundation Conference: Isotopes in Biochemistry, Philadelphia, Blakiston, 1951.

[20] Warner, G F, Sweet, N J., and Dobson, E. L.: Sodium space and body sodium content exchangeable with sodium^{24} in normal individuals and patients with ascites, Circulation Res 1 486-490, 1953

[21] Heidelberger, C., and Weiss, S M.: The distribution of radioactivity in mice following administration of 3,4-benzpyrene-5-C^{14} and 1,2,5,6-dibenzanthracene-9,10-C^{14}, Cancer Res 11 885-891, 1951.

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would produce physiologic changes due to radioactivity. Beyond this level, the harmful effects produced by ionizing radiations in living organisms become visible in the form of clinical and pathologic changes, grouped under the term of "radiation-effects."[10]

A man in contact with radioactive material is confronted with two dangers; he may be exposed to excessive amounts of external radiation, or he may absorb radioactive compounds through the nose (inhalation of dusts and vapors), the skin (contamination of intact or broken skin surfaces) and the mouth (ingestion).[11] In any event, the result of excessive exposure to radiation is a shortening of life. After very large doses, death may take place within a few days or sometimes hours, whereas, when the radiation dose is decreased, death is brought about by more insidious and chronic injuries. Therefore, we shall try to distinguish between acute and chronic effects.

The acute effects of massive doses of radiation on man were fully illustrated by the many casualties that occurred when atomic bombs were dropped on Hiroshima and Nagasaki. Lesions of the gastro-intestinal tract and of the hemopoietic system were mainly responsible for the acute radiation deaths.[12] Similar devastating effects are a hazard of atomic energy plants and laboratories handling radioactive material, but in ordinary circumstances the chronic effects of ionizing radiations are the most frequently observed and the most important to the practicing physician. Among these, genetic changes are probably the most insidious and the most undesirable, but their consideration is beyond the scope of this brief review, and for further information the reader is referred to the recent report of the Committee on Genetic Effects of Atomic Radiation[13] Radiations also cause tissue changes resembling those of aging and usually have a striking and depressing effect on the hemopoietic system. Plutonium and radioactive cerium (Ce^{144}) accumulate in the liver, where they can induce an acute yellow atrophy of the organ.[14] By far more interesting, however, is the recognized fact that ionizing radiations can produce in man and experimental animals a variety of malignant tumors.[15] In fact, only 15 years had elapsed since Roentgen's discovery, when Hesse published his monograph entitled: "The Symptomatology, Pathogenesis and Therapy of Roentgen-Ray Carcinoma," in which he described 54 patients with carcinoma of the skin resulting from overexposure to x-rays. Many other instances of radiation-induced tumors were recorded after Hesse's initial observation: briefly, we can mention the carcinoma of the lung so frequent in the miners of the uranium-bearing ores of Schneeberg and Joachimsthal in Central Europe, the high incidence of leukemia in radiologists and among the survivors of Hiroshima and Nagasaki who were exposed to sublethal doses of radiation, the visceral sarcomas and carcinomas produced by the injection of Thorotrast and the many kinds of tumors induced by a variety of radioactive isotopes in experimental animals. Strictly pertinent to the field of orthopedic surgery are the lesions induced in dial painters by radium-mesothorium.[16] These dial painters, it will be recalled, used a self-luminous paint containing zinc pyrosulphate and small amounts of radium, mesothorium and radiothorium. In the early years of this industrial process many dial painters succumbed to a progressive and often fatal illness, resulting from the ingestion of considerable amounts of this paint through the notorious practice of tipping the brushes with their lips. The lesions were characterized by rarefaction, necrosis and fractures of bones (referred to as "radiation osteitis") combined

[10] United Nations Report of the United Nations Scientific Committee on the Effects of Atomic Radiation, Supplement No 17(A/3838), New York, 1958

[11] Lisco, H Potential Hazards and Pathological Aspects of Radioactive Isotopes, Proc. Ninth International Congress on Industrial Medicine, September, 1948.

[12] Liebow, A A, Warren, S, and DeCoursey, E.: Pathology of atomic bomb casualties, Am J. Path 25:853-1027, 1949

[13] Committee on Genetic Effects of Atomic Radiation: Genetic effects of atomic radiation, Science 123:1157-1164, 1956.

[14] Brues, A. M Toxicity of radioactive isotopes *in* Glasser's Medical Physics, vol 2, p 465, Chicago, Year Book Pub, 1944.

[15] Lisco, H · Radiation and Carcinogenesis, International Forum, vol 2, January, 1954

[16] ———: Bone as a critical organ for the deposition of radioactive materials *in* Ciba Foundation Symposium on Bone Structure and Metabolism, pp 272-282, London, Churchill, 1956

with severe damage of the bone marrow. In more chronic cases, the skeletal lesions appeared first as discrete or confluent areas of bone destruction and rarefaction, which were slowly progressive and were complicated in several instances by malignancies developing in the damaged skeleton. Similar lesions have been observed in a number of patients who, in the 2nd and the 3rd decades of this century, were treated with radium salts for a great variety of illnesses. Thus, radium poisoning, although a rare disease, is now a well-recognized clinical and pathologic entity, which the physician should always consider in the investigation of a disease. In this connection, it is of special interest to know that heavy elements, such as uranium, plutonium and other transuranic elements of the actinide series, as well as radioactive isotopes which are formed in abundance in the fission of uranium or plutonium, are deposited and fixed in the skeleton. Though bone sarcomas have not been reported yet in man, tumor formation in bone following the administration of plutonium and of various fission products, notably strontium (Sr^{89}), has been reported in experimental animals and has suggested the possibility that these bone-seeking isotopes may produce similar harmful effects in man.[17]

RADIOACTIVE ISOTOPES AS TRACERS

A tremendous volume of literature has accumulated in the past few years over the use of radioactive isotopes as tracers in biochemical research.[18, 19] Briefly, in this type of investigation, one takes advantage of the radioactivity of a compound to follow its distribution, concentration and fate in the animal body. Most of the artificial radioactive isotopes can be used for such purpose, but the ones most frequently employed are Deuterium (H^2), $Carbon^{14}$, $Nitrogen^{15}$, Na^{24}, P^{32}, S^{34} and S^{35}, Fe^{55}, Co^{60}, Cu^{64}, Zn^{65} and I^{131}. The types of biochemical problems that are best studied by the use of tracers are essentially three: (1) anatomic distribution of a compound; (2) precursor-product relationships; and (3) analysis of rates of processes.

By administering isotopic material to the animal body, it is possible, by subsequent analysis, to ascertain its distribution in the various tissues or products and, by analogy, the distribution of its stable counterpart. The localization of the isotopic atom can even be established at the cellular level by the use of the technic of autoradiography, mentioned above. This type of investigation has been particularly useful in determining the distribution in the body of administered compounds and/or their products, such as water, electrolytes,[20] carcinogenic hydrocarbons,[21] and others.

The purpose of experiments dealing with the precursor-product relationship is to determine whether compound A can be converted into B in the organism. This can be accomplished by analyzing for isotope the carefully purified compound B after administration of isotopic compound A. This technic has given dramatic results in the investigation of the biosynthesis of various components of the animal body, such as steroids, amino acids, nucleic acids and hemoglobin.[22] Conversely, the same technic applies to the study of the catabolism of these compounds and/or others, such as carbohydrates and fatty acids.

Finally, from the use of isotopes in the analysis of rates of processes has evolved the modern concept of continuous turnover (synthesis, degradation and replacement) of certain body constituents (proteins, depot lipids, bone minerals, etc.) hitherto considered to be relatively stable in their composition.

These different technics are naturally applicable to the investigation of bone metab-

[17] Lisco, H · Bone as a critical organ for the deposition of radioactive materials *in* Ciba Foundation Symposium on Bone Structure and Metabolism, pp 272-282, London, Churchill, 1956

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[16] ———: Bone as a critical organ for the deposition of radioactive materials *in* Ciba Foundation Symposium on Bone Structure and Metabolism. pp 272-282, London, Churchill, 1956.

roidism and in a small percentage of thyroid carcinomas.[31,32] Most diagnostic technics with isotopes are essentially tracer studies, wherein the radioactive atoms follow the same metabolic pathways as do their stable counterparts in the body. For further information the reader is referred to the "Table of radioactive isotopes of value in diagnostic and tracer studies," recently compiled by Belcher and Mayneord.[33]

In orthopaedic surgery,[34] the use of isotopes as diagnostic and therapeutic tools is still limited mainly to two isotopes—P^{32} and I^{131}. P^{32} was used in the early days of radioisotope work in an attempt to treat prostatic and mammary cancers metastatic to bone. Though results were not outstanding, further studies showed that P^{32} produced some relief of pain and occasional evidence of healing, and now it is agreed that this type of therapy may have a small place in the palliation of cancer metastatic to bone. Sr^{89}, which is deposited selectively in the skeleton, was found to concentrate in regions of active new bone formation, such as an osteogenic sarcoma or osteoplastic metastases from a prostatic carcinoma. In a few cases, Sr^{89} displayed an inhibitory action on metastases, while the serum acid phosphatase dropped to normal levels. However, its use was discontinued because of the concomitant severe marrow depression. Radioactive gallium (Ga^{72}) was also found to concentrate in areas of osteoplastic metastases from soft-tissue carcinomas, but it produced undesirable radiation effects at subtherapeutic dosage levels. For the same reason, the use of radiosulfate in the treatment of chondrosarcomas was found to be impractical, though S^{35} concentrates in the growing portions of chondrosarcomas.[35]

[31] Rosenthal, D J, and Lawrence, J H Radioactive isotopes in medicine, Ann Rev Med 8 361-388, 1957

[32] Garland, L H, and Heald, J H Radioactive isotopes in the diagnosis and treatment of cancer, J A M A 164 1096-1098, 1957.

[33] Belcher, E H, and Mayneord, W V: Radioactive isotopes in medical diagnosis Progress in nuclear energy Series VII. Medical Sc 1·1-32, 1956.

[34] Lawrence, J H., and Tobias, C A Radioactive isotopes and nuclear radiations in the treatment of cancer, Cancer Res 16 185-193, 1956

[35] Beierwaltes, W. H, Johnson, P. C., and Solari, A J · Clinical Use of Radioisotopes, Philadelphia, Saunders, 1957.

Radioiodine (I^{131}) is generally considered to be of value in a small percentage of metastatic thyroid carcinomas. The therapeutic effects of radioiodine depend upon its selective concentration in thyroid tissues, but unfortunately only a small percentage of cases has spontaneous concentration of I^{131} in the metastases. It was thought, at first, that radioiodine would concentrate in larger quantities in well-differentiated tumors, but Maloof and co-workers[36] are of the opinion that predictions based on the microscopic picture are frequently wrong, and furthermore that the distribution of radioiodine is unequal within a single metastasis. The uptake of I^{131} in bone metastases may be increased by thyroidectomy or by administration of propylthiouracil or thyroid-stimulating hormones; but, even so, one is faced with the danger of stimulating thyroid metastases without achieving a corresponding increase in function. Nonetheless, the results presented by Maloof and co-workers are not disheartening, for out of 21 patients, I^{131} produced radiologic disappearance of bone metastases in 2, a slight decrease in size in 3, a retarded growth in 4, and a relief of pain in 5. The introduction of the whole-body scintillation scanner, which permits the visualization of radioisotopes distribution in vivo, is expected to improve the efficiency of this kind of therapy through more accurate localization of metastases.[37]

To the field of therapeutic uses belongs also an important problem of this atomic age, that is the mobilization of radioisotopes that have been deposited in bones. As mentioned above, there is a group of radioisotopes which have a tendency to accumulate in bones and therefore have been called "bone-seekers." In the adult, "bone-seekers" are deposited principally in the newly forming haversian systems, where they become entrapped and isolated from contact with circulating fluid. Once incorporated in bone, regardless of the manner of initial deposition, most radioactive materials remain there for long periods of time. As it is likely that man, through radioactive fall-out and

[36] Maloof, F, Vickery, A. L, and Rapp, B.: An evaluation of various factors influencing the treatment of metastatic thyroid carcinoma with I^{131} J. Clin Endocrinol 16·1-27, 1956.

[37] Lawrence, J H, and Tobias, C. A.: Radioactive isotopes and nuclear radiations in the treatment of cancer, Cancer Res 16:185-193, 1956.

olism. Elements which have an affinity for the skeleton and therefore can be used for this purpose are divided into two categories.[23]

One group of elements (americium, plutonium, yttrium, barium, zirconium, cerium, gallium, niobium, lanthanum, praseodymium, neodymium, promethium, samarium, eruopium, neptunium, actinium) have been shown to concentrate specifically in the organic or osteoid matrix of forming bone. Shortly after deposition, one member of the group may displace another, but usually the deposited material remains for the entire life of the bone, *with scarcely any decrease in concentration.* Bone matrix formation has also been investigated by using S^{35}-labeled chondroitin sulfate; it was found that calcification of the osteoid matrix is associated with an accelerated synthesis and removal of chondroitin sulfate in the adjoining cartilage.[24]

Another group of elements (calcium, strontium, uranium, radium, magnesium, lead) are deposited mainly in the mineral phase of bone. However, the 3 isotopes most frequently used in studies of mineral metabolism are Ca^{45}, P^{32} and Sr^{89}. All 3 are beta emitters.[25] Sr^{89}, though not a physiologic compound, has been shown, in low concentration, to parallel closely the metabolism of calcium, while P^{32} accumulates in bone in quantities larger than in any other organ, though sizable amounts are found in other tissues. Ca ions, administered by the intravenous route, are found within minutes *in bone, a fair proportion of these ions being in* the exchangeable fraction. After a few hours, a large proportion of Ca ions are found in the nonexchangeable fraction; and after several hours, the addition of Ca to the skeleton can be accounted for only by new bone deposition. In young, growing animals as well as in adults, discrete areas of radioactivity after injection of radioisotopes (hot-spots) can be correlated with areas of bone growth and reconstruction, such as an epiphysis or a callus. P^{32} also accumulates more readily in the epiphyses than in the diaphyses of bone. In rachitic individuals, the rate of P^{32} deposition is decreased, but it is restored to normal by vitamin D, which has no effect on normal animals Strontium[90], a new radioisotope of strontium recently made available, has been shown to follow the same pattern of Sr^{89} and Calcium.[26]

The absorption and the excretion of calcium and phosphate were also the object of tracer studies.[27] The metabolism of these elements was shown to be essentially similar. Much of *the calcium and the phosphate of the feces* was demonstrated to derive from unabsorbed food material, while the absorbed calcium and phosphate in excess of that retained by the skeleton was excreted for the most part by the way of urine.

So far, radioisotopes have been confined to the study of the physiology of bone, but it is obvious that a vast field is open for further investigation by the tracer technic of pathologic conditions, such as the healing of bone fracture and the fate of bone grafts.[28, 29, 30]

RADIOISOTOPES IN THE DIAGNOSIS AND THE TREATMENT OF DISEASES

Whereas the use of radioactive isotopes as diagnostic tools has been quite successful and is very likely to become more and more popular, it should be stated clearly that their use *as therapeutic agents, on the* whole, has been rather disappointing, only two actually providing important therapeutic applications: P^{32} in polycythemia vera and I^{131} in hyperthy-

[23] Comar, C L., and Wasserman, R. H Radioisotopes in the study of mineral metabolism Progress in nuclear energy. Section VI Biological Sc 1 153-196, 1956.

[24] Ross, J F: The diagnosis and investigation of disease with radioactive isotopes Progress in nuclear energy. Series *VII. Medical Sc 1:33-48, 1956*

[25] Greenberg, D. M. Tracer Studies on the Metab-

[26] Ray, R. D, Thomson, D. M, Norvelle, K. W., and Laviolette, D.: Bone metabolism. II. Toxicity and metabolism of radioactive strontium (Sr^{90}) in rats, J. Bone & Joint Surg 38A:160-174, 1956

[27] Bauer, G C. H., and Carlsson, A: Metabolism of bone salt investigated by simultaneous administration of ^{45}Ca and ^{32}P to rats, J. Bone & Joint Surg 37B 658-662, 1955.

[28] Ross, J F.: The diagnosis and investigation of disease with radioactive isotopes Progress in nuclear energy Series VII. Medical Sc. 1:33-48, 1956.

[29] Cohen, J., Maletskos, C. J., Marshall, J. H, and Williams, J B.: Radioactive calcium tracer studies in bone grafts, *J. Bone & Joint Surg.* 39A:561-577, 1957.

[30] MacDonald, N. S, Lorick, P. C., and Petriello, L. I.: Healing bone fractures and simultaneous administration of radioisotopes of sulfur, calcium and yttrium, Am. J. Physiol. 191 (1):185-188, 1957.

30

Amputations

The loss of any portion of a limb disables the individual to a variable degree, depending on the extent of loss, the age of the patient, the propriety of surgical attack, and the post-operative management. One or all of these factors may be operative when the patient cannot be restored to a useful existence

INDICATIONS FOR AMPUTATION

1. Destruction of limb as by crushing injury
2. Loss of blood supply—gangrene
3. Irremediable pain of circulatory origin
4. Malignant tumor with hope of eradication
5. Cosmetic reason
6. Uncontrolled infection

PREOPERATIVE PREPARATION

Although the procedure may be lifesaving, haste is rarely indicated The patient must be brought to the best possible condition to withstand and survive the surgical trauma. Appropriate measures are instituted to control diabetes, dehydration, anemia, shock, infection and cardiac insufficiency. If the limb is gangrenous and/or infected, refrigeration of the affected part will reduce absorption of toxic products and improve the condition of the patient while awaiting surgery. The amazing response to reducing the temperature of the part is noted in the rapid subsidence of the fever, and the extremely sick patient almost at once becomes rational and comfortable. Caution should be exercised regarding the proper positioning of the ice. Very low temperatures destroy the small caliber vessel walls and, following removal of the cold, permits extravasation of serum and perivascular edema, hemoconcentration and intravascular obstruction. In consequence, the tissues are devascularized and heal poorly Therefore, it is important to keep the upper level of the ice well away from the contemplated site of amputation. Refrigeration is superior in arresting the spread of gangrene and infection, improving the general condition, alleviating the pain, and minimizing the shock of surgery. It may be applied for an indefinite period but as a rule is instituted about 24 hours before surgery.

TYPES OF AMPUTATION

The open type is one in which the stump is left open for drainage and is always done in the presence of established or potential infection. The resulting part represents a cross section of the extremity. It can be done quickly with a minimum of shock. An open amputation done with flaps prepared for early closure is termed an *open flap* amputation; without flaps, an *open circular* amputation.

TECHNIC FOR OPEN CIRCULAR AMPUTATION

This is the safest procedure. One aims at producing a slightly concave, open cross section with the skin slightly longer than the superficial muscles and the deep muscle slightly shorter than the overlying muscle. A circular incision is made only through the skin which retracts. The fascial incision is also circular and at the level of skin retraction. The superficial muscle is cut at the edge of the fascia and retracts. The deep muscle is cut at the level of superficial muscle retraction. Periosteum is cut at the level of deep muscle retraction. The nerves are pulled down gently, cut, and allowed to retract into the fascial planes Excessive traction, crushing, or alcohol injection predisposes to neuroma formation. Large nerves are transfixed and ligated above the point of section to guard against bleeding. Large vessels are transfixed.

The amputation site is at the site of a fracture in the case of a compound infected frac-

industrial hazard, will become increasingly more exposed to bone-seeking isotopes, and as some of these isotopes have been demonstrated to be carcinogenic in man and experimental animals, the problem of their mobilization from skeletal deposits is of considerable importance.

Once the living organism has been exposed to a dangerous concentration of bone-seeking radioisotopes, different procedures can be attempted in order to minimize the consequences. These procedures can be divided into 4 groups:[38] (1) minimization of absorption; (2) interference with calcium metabolism; (3) use of chelates and other complexing agents; and (4) miscellaneous approaches.

Minimization of absorption may be effective in those cases in which the radioisotopes have been swallowed. The treatment, with emetics, precipitating agents and cathartics, must be started immediately, as soluble radioisotopes are absorbed rapidly in the gastrointestinal tract. Clearly, this treatment is of no use in those cases in which the "bone-seekers" are inhaled through the lungs, where the rapidity of absorption is nearly equivalent to that of an intravenous injection.

Once the radioisotopes have been absorbed and deposited, some mobilization may be effected by interference with calcium metabolism. For example, a low calcium diet plus ammonium chloride, thyroid extracts and parathyroid extracts may increase the excretion of radium, strontium and calcium, though at such a slow rate that the benefits are rather limited. Decalcification therapies have no effect whatsoever on the excretion rates of plutonium and rare earth elements.

The use of chelating and other complexing agents is based on the principle that chelated and complexing agents remove the metal ion (such as strontium or plutonium) from combination with cellular constituents, with subsequent elimination of the complexed metal (metal ion plus chelating agent) from the body. At present, the 4 principal chelating agents are: EDTA (ethylenediaminetetraacetic acid), BAL (2, 3-dimercaptopropanol, British anti-Lewisite), sodium citrate and ATA (aurintricarboxylic acid).[39] Studies with man and animals have shown that these agents are significantly effective in removing from tissues previously deposited radioisotopes of plutonium, lanthanum, yttrium, polonium, thorium, strontium and beryllium.

Other approaches, grouped under the term of miscellaneous approaches, include (1) the use of colloidal carriers and zirconium salts; (2) disturbance of the organic matrix of bone by induction of scurvy; (3) use of protein-free diet to disturb bone proteins; (4) causation of bone resorption with subsequent use of a diet calculated to restore the bones; and (5) use of endocrine changes, such as induction of hypothyroidism. The value of these methods has not yet been fully established

[38] Comar, C L., and Wasserman, R H: Radioisotopes in the study of mineral metabolism. Progress in nuclear energy. Section VI. Biological Sc 1:33-48, 1956.

[39] Lindenbaum, A., and Lisco, H.: Autoradiographic study of localization of aurintricarboxylic acid in experimental beryllium poisoning, Proc. Soc. Exper Biol & Med 92:354-357, 1956.

SITES OF AMPUTATION

This is determined by (1) the type of prosthesis; (2) function of the part; (3) muscle balance; (4) and adequacy of the circulation as determined by oscillometric readings, histamine flare tests and pulsations.

Through the Foot

The main factors to consider are good thick skin coverage to withstand trauma, and the preservation of muscle, because imbalance results in a disabling deformity

1. **Amputation of the Great Toe.** Does not affect walking at the normal pace The loss of "take-off" is apparent only with rapid gait.

2. **Amputation of the Second Toe.** Results in hallux valgus.

3. **Amputation of the Fifth Toe.** No disturbance results; preferred to plastic procedures for overriding deformity of the toe

4. **Amputations of All the Toes.** Little disturbance in slow walking A sponge-rubber insert in the toe of the shoe and a spring steel band running longitudinally in the sole makes an excellent support and provides the necessary fulcrum for the "take-off "

5. **Amputation Through the Metatarsals.** No deformity. Very satisfactory.

6. ***Proximal to Metatarsal Bases.*** *Inversion* deformity results.

7 **Tarsals Proximal to Anterior Tibial Insertion.** Severe equinovalgus will result.

8. **Syme Amputation.** This is done through the ankle, and the heel flap is brought over the lower end of the tibia. This is an end-bearing stump with superior function. Skin accustomed to weight-bearing covers the end. However, a large stump end results and necessitates a bulky prosthesis, which consists of a molded leather socket with supporting steel on each side fixed to a footpiece. An advantage is that the patient can walk very well without the prosthesis. A disadvantage is that it is cosmetically unsuitable for a woman to wear such a prosthesis. The blood supply of the heel flap originates from the medial side of the ankle through a branch of the posterior tibial artery. Therefore, disarticulation of the ankle must be done from the inside out, removing the os calcis subperiosteally. This reduces the danger of skin necrosis. The skin

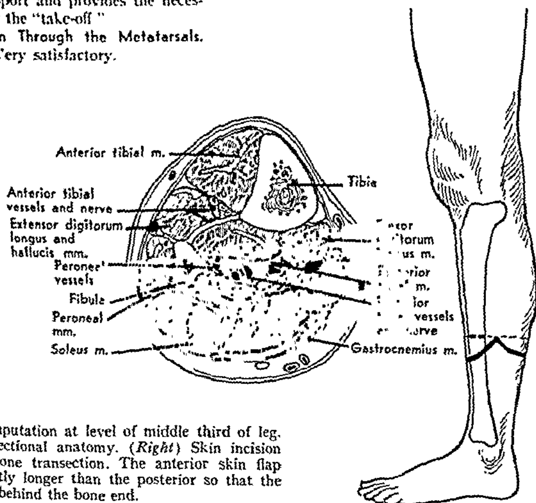


Fig. 540 Amputation at level of middle third of leg. (*Left*) Cross sectional anatomy. (*Right*) Skin incision and level of bone transection. The anterior skin flap should be slightly longer than the posterior so that the suture line lies behind the bone end.

ture. Otherwise, it is done as far distally as viability will allow.

Skin traction is applied immediately postoperatively and continuously. First, the stump is coated with skin adherent. Stockinette is applied smoothly and evenly and is extended a distance beyond the stump. To the distal end is attached the traction weight. The weight is removed daily, and the stockinette is rolled back for daily dressings. Eventually, the skin is pulled down, the bone end becomes covered with granulations, and the skin margin is closed by scar contracture. Without skin traction, the skin and the muscles recede, and reamputation at a higher level is necessary. Closure of the open amputation and preparation for the prosthesis is done from 6 to 8 weeks after the first stage. The scar is excised with a wafer from the end of the bone. The muscle and the fascia are found to be firmly attached to the distal end of the bone, and it is unnecessary to expose them. The skin is undermined a distance of 10 cm. and closed. Skin traction postoperatively relieves tension on the suture line. The fibula is cut off shorter than the tibia. Instead of this stage, reamputation and primary closure may be done at a more ideal site.

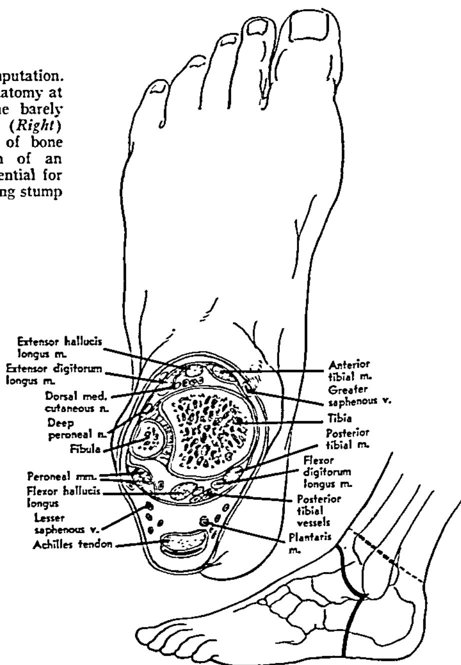


FIG. 539. The Syme amputation. (*Left*) Cross sectional anatomy at the level of the sawline barely above the ankle joint. (*Right*) Skin incision and level of bone transection. Preservation of an adequate heel flap is essential for covering the weight-bearing stump end.

As much femur as possible should be preserved, because the difficulty in prosthetic fitting is proportionate to the shortness of the stump.

Types

1. *Tendoplastic.* The patella is removed, and the tendinous portion of the quadriceps is brought over the end of the femur.

2. *Gritti-Stokes.* The patella is placed over the end of the sectioned femur. The possibility of infection and nonunion should be considered.

A higher level of amputation is necessary when circulation is poor. The junction of the middle and the lower thirds is the desirable site. Only skin and fascia should cover the end of the femur. Redundant muscle must be coned back because it (1) turns to avascular scar, (2) slips about on the end of the bone, (3) becomes edematous and (4) makes prosthesis-fitting difficult. A minimum of 4 or 5 inches distance from the knee provides the necessary clearance for provision of an artificial joint. In shorter stumps muscle balance is poor.

Hip

Disarticulation is almost never indicated. Even a small stump of bone can be left and is useful for grasp by the contours of the socket which is shaped like a quarter hemisphere. A smooth contour of the hip is useless for controlling the limb.

Hindquarter Disarticulation

This is the removal of the entire lower extremity and the corresponding half of the pelvis. It is an exceedingly shocking operation and should be done only for malignancy with some hope of eradication.

Fingers

As much length as possible should be conserved. The tip should be covered with a thick

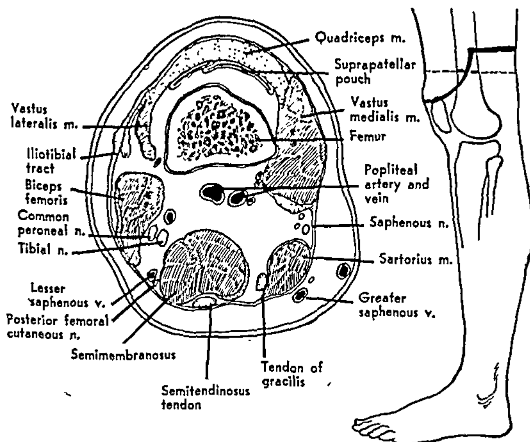


Fig. 542. End-bearing amputation through the lower third of the thigh. (*Left*) Cross sectional anatomy. (*Right*) Skin incision. The anterior flap is made a little longer than the posterior. When it is desired that the bone end be covered by the quadriceps aponeurosis (tendoplastic amputation of Kirk), the anterior flap is cut deeply so as to contain the quadriceps tendon.

flap postoperatively tends to displace to the medial side. Adhesive fixation for a few weeks prevents this.

Through the Leg

The *ideal length* is 5 to 7 inches of tibia, varying with the height of the individual Excessively long stumps are difficult to fit with a prosthesis, the prosthesis contour is disproportionate, and the inadequate circulation distally results in pain and ulceration Shorter stumps are still very valuable, because it moves the knee of the prosthesis, and better control of the limb is obtained in climbing stairs or descending inclines. However, prosthesis-fitting is difficult. By sectioning the hamstrings and removing the head of the fibula, fitting is easier. Extremely difficult cases may be fitted by bending the stump to 90° and fitting an end-bearing prosthesis. *The fibula should be cut off 3 cm. above the end of the tibia. A long fibula results in excessive pressure by the prosthesis* and leads to a painful bursitis over its tip and a sprung and painful upper tibiofibular joint. Removal of the entire fibula is permissible if a small unstable piece remains or to aid skin closure. Prosthesis-fitting becomes difficult when the fibula is absent. Weight-bearing is distributed on the upper flare of the tibia. If the stump is too short, the thigh corset may be extended so that additional support on the ischial tuberosity is obtained.

Through the Thigh

The supracondylar area is the ideal site, and a conical end-bearing stump is produced.

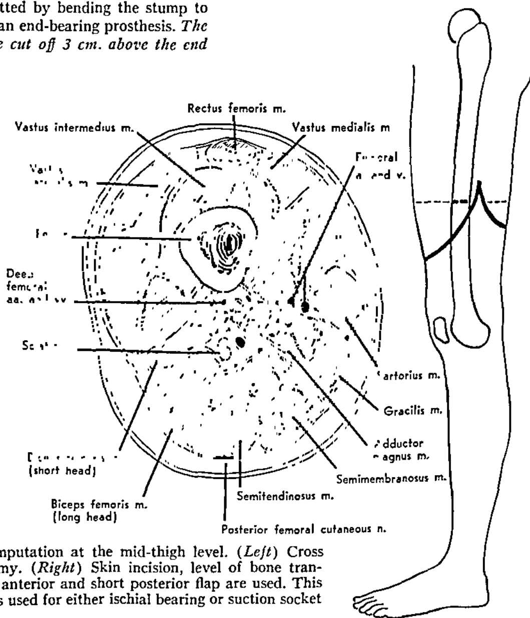


Fig. 541. Amputation at the mid-thigh level. (*Left*) Cross sectional anatomy. (*Right*) Skin incision, level of bone transection A long anterior and short posterior flap are used. This type of stump is used for either ischial bearing or suction socket prostheses.

covering the end, (2) no protruding bony prominences, (3) the radio-ulnar joint is intact, and (4) vascularity and sensation of the stump are unimpaired.

Cineplastic Procedure

This consists of surgically constructing a canal transversely through a muscle. Into this canal is inserted a rod attached to a flexing

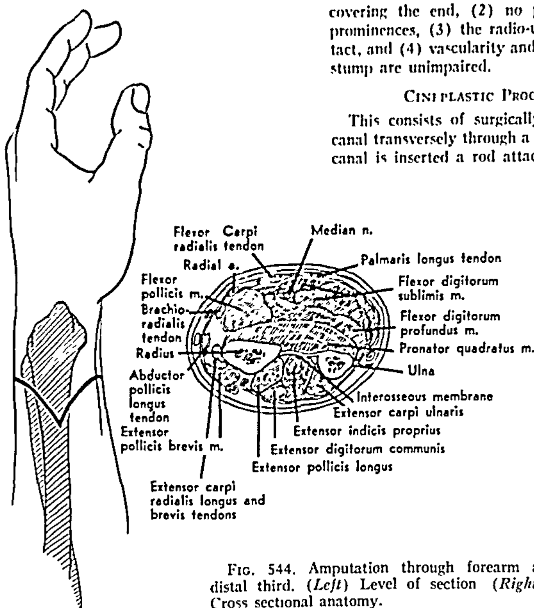


Fig. 544. Amputation through forearm at distal third. (*Left*) Level of section (*Right*) Cross sectional anatomy.

All possible length is preserved, by orthopaedic reconstruction if necessary. If the thumb is short, deepening the web between the index finger and the thumb improves the grasp. In the absence of the thumb, transplanting the index finger to the thumb has occasionally been successful

Removal of All Fingers and the Thumb. This results in a "mitten hand." By excising the 2nd and the 3rd metacarpals and the capitate bone, a bifid stump is created for useful pinching action.

Wrist

Disarticulation at this level is advantageous because the prosthesis needs no above-the-elbow cuff and is easily used; and supination and pronation movements are retained. The requirements are (1) adequate palmar skin

mechanism in the artificial hand. Contraction of the muscle is effective as a motor in producing flexion of the fingers. The operation is used most commonly in the forearm.

Forearm

The distal third is unsuitable as the amputation site because the circulation is poor. The underlying tissues are largely tendinous, the skin is thin, and the stumps become cold and cyanotic The ideal level is the junction of the middle and the lower thirds. The stump is smoothly tapered, the scar is terminal, rotation is preserved, and cineplastic procedures can be performed. As much length as possible should be preserved in order to retain elbow motion In a short stump, removal of the distal 1 inch of the biceps tendon will facilitate fitting of the prosthesis.

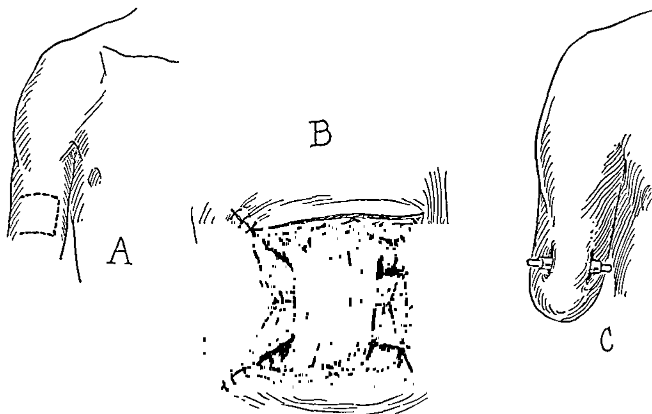


FIG. 543. Cineplastic amputation. (A) Outline of pedicle flap. (B) The flap is sewed in the form of a tube and inserted transversely through the muscle. A skin graft covers the surface area. (C) Peg inserted in tunnel. Cables attach to protruding ends of pég.

pad (full-thickness graft or flap is necessary) in order to withstand the constant trauma. Fingernails facilitate the pinching action, and even the smallest portion retained is useful. The thumb is the most important in grasp and pinch. The index and the middle fingers, next in importance, are the strong stabilizing units that work with the thumb. The ring and the little fingers are least important and provide mobility to the metacarpal arch by their wide range of motion at the carpometacarpal articulations. An interphalangeal disarticulation is acceptable if the bulky head of the phalanx is rounded off.

Index Finger. This is important in grasping and pinching. An *amputation above the proximal interphalangeal joint results in loss of pinch. Therefore, amputate at the base or disarticulate at the metacarpophalangeal joint* so that the stump will not impede the pinching action of the middle finger If amputation is required above this level, all but the base of the metacarpal should be removed, as the unstable metacarpal is an obstacle to grasp.

Index and Middle Fingers. This is done through the metacarpals. Resection is done at the base of the metacarpals, fashioning a web between the remaining fingers and the thumb. The web should be sutured by the zigzag method to prevent the formation of contracture.

Index, Middle and Ring Fingers. This is done with a web space that drops in a gradual arc from the 5th finger to the metacarpal head of the thumb. The protruding heads of bone in this arc should be resected

Rotation of the index or the middle finger during flexion follows amputation through the 3rd or the 4th metacarpal because the stabilizing action of the transverse metacarpal ligament on the palmar arch is lost. To avoid this, *the entire metacarpal in the amputated segment must be removed* so that the adjacent metacarpal heads are collapsed together. Less commonly used methods are transplantation of an adjacent metacarpal to the site of the amputated one, or supplying a bone graft.

Little Finger. As much metacarpal as possible is preserved. The stability of the palm is thereby maintained, and the attachment of the intrinsic muscles is preserved.

Thumb. This is the most important digit.

and pressure area about the ischial tuberosity.

3. *Thigh.* Flaps equal, scar falls just behind the bone; or a long anterior flap so scar will be well above the bond end.
4. *End-bearing Stumps at Distal End of Femur.* Long anterior and short posterior flap.
5. *Below the Knee.* Anterior and posterior flaps of equal length allow scar to lie transversely immediately behind the tibia. Avoid scar on side of the stump.
6. *Syme Amputation.* Long posterior heel flap.

Muscles divided just distal to the saw line, the muscle retracting to the bone level. Do not separate the skin from the tendon or the muscle when the configuration of both is identical, because this aids in grouping the muscles about the side of the bone. Bulky muscle masses should not be placed over the bone end.

Periosteum is cut at the bone level. The bone is cut transversely except in end-bearing amputations where bone is sectioned so that the end lies parallel with the ground while the patient is in the standing position. The bony edges are rounded off with a file Bone dust is irrigated out.

Nerves are isolated, pulled down gently, sectioned and allowed to retract. Strong tugging at the nerve predisposes to neuromata and phantom limb pains. Avoid injection, closure, or burying nerve in the bone.

Major vessels are doubly ligated with plain catgut. Minor vessels are singly ligated. Then the tourniquet is released, and hemostasis is completed.

Skin flaps are approximated with interrupted skin sutures. No tension sutures are used.

A drain is placed in the wound and removed in 48 to 72 hours.

An elastic compression bandage is applied smoothly. This minimizes edema, eliminates dead space and molds the stump

Absolute bed rest with the limb elevated until healing is complete.

Plaster splints may be used to prevent flexion contracture of knee

Sutures are removed at 14th day.

Exercises are started.

Elastic bandaging until maximum shrinkage has taken place

Limb is fitted—minimum of 8 to 12 weeks after surgery.

AMPUTATION IN THE PRESENCE OF PERIPHERAL VASCULAR DISEASE

Indications

Gangrene, uncontrolled infection and ischemic pain are definite indications for amputation.

Site

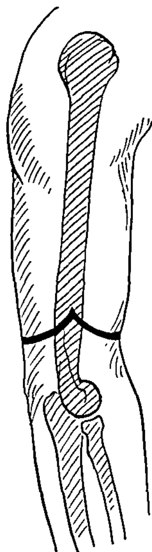
The site is determined by adequacy of circulation which ensures healing of the stump, guards against infection and resists the pressure and the friction of the limb socket.

Gangrene of the Foot

If the dorsalis pedis and/or the posterior tibial pulses are present, amputation may be done in the viable tissue immediately adjacent to the necrotic area. If infected, open amputation followed by secondary closure is indicated. If foot pulsations are absent, and popliteal pulse is good, the case being devoid of infection, amputation through the leg may be attempted. To ensure the adequacy of circulation, histamine flare and oscillometric reading should be done. In the presence of infection, amputation at a distance is mandatory whether or not popliteal pulses are present.

End-Bearing Stump

An end-bearing stump should be avoided because pressure will reduce the circulation of an already poorly nourished tissue. A gangrenous toe in an arteriosclerotic, particularly a diabetic, even though a successful distal amputation is a possibility, is a warning of impending disaster. Gangrene and uncontrollable infection in the immediate future are almost a certainty. Success of rehabilitation depends in large measure upon seizing the early opportunity of performing an amputation through the leg. At this stage healing of the stump, toughening of the skin and adaptation to the socket are easier to obtain. Delay, overcautious conservatism, and unwillingness on the part of the patient and his family to accept the inevitable, have resulted in totally disabled bilateral above-knee am-



Shoulder Disarticulation

Removal of the head of the humerus is the only indication for this operation.

Forequarter Amputation

Performed for malignant growths.

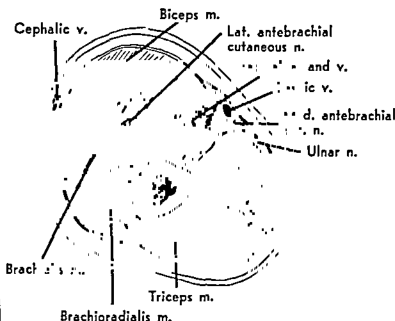


Fig. 545. Amputation through supracondylar area of arm. (*Left*) Level of section. (*Right*) Cross section.

Forearm Prosthesis. This consists of a plastic shell which fits over the forearm The sides are supported by steel bars which pass upward to elbow joints which are fixed to an upper arm cuff. Distally, the bars are attached to a socket in which the artificial hand or hook is mounted. Activation of the latter is by a cable extending to a cuff about the opposite shoulder.

Elbow

Disarticulation should not be done, because the end of the humerus is too bulbous to allow proper fitting of a prosthesis.

Upper Arm

The supracondylar region is the ideal site. As much length as possible should be saved, because the strength is proportional to the size. Also, a stump of 2 inches or less is of no functional value but should be preserved to maintain the contour of the shoulder.

PRINCIPLES OF AMPUTATION TECHNIC (SLOCUM)

Clean Operative Field. Fresh wounds are cleansed and débrided. Old wounds should display a cherry-red granulating surface.

Tourniquet. Except in vascular disease

Good skin covering over the end of the stump. Normal sensation and mobility and adequate subcutaneous fat provide protection against trauma. The scar is placed away from pressure and tension. Suturing without tension and avoiding redundancy is mandatory.

Length of the flap:

1. *Upper extremity*
   Above the wrist. Anterior and posterior flaps of equal length
   Below the wrist. Long anterior and short posterior flap utilizes the tough palmar skin over the end.
2. *Hip.* Racquet incision, scar anterior and lateral away from fecal contamination

A short limb is not effective as a lever in moving the prosthesis and frequently falls out of the socket. It is prone to contracture of the adjacent joint, is difficult to fit, and frequently reamputation is done. A longer stump likewise is inadequate because the excessive up-and-down motion within the socket causes breakdown of the suture line. It is difficult to fit to a prosthesis. However, *the stump in children should be made lengthy because growth lags behind in the amputated extremity* and eventually will be suitable for fitting. The length of bone is determined by the presence of tissues suitable for protection of the bone, attainment of an effective lever arm to activate the limb, and suitability for prosthetic fitting.

An ideal stump is cylindrical and should taper gradually to a smoothly rounded end. Deviations from this, because of projecting bone and massive redundant muscle or skin, make fitting difficult, cause breakdown and are productive of pain. One must remember that in children the rate of growth of bone exceeds that of the soft tissue. This should be compensated for at surgery by leaving a slightly longer muscle and greater skin redundancy. The fibula outgrows the tibia; therefore, a sufficient length of fibula should be removed in amputation through the leg. Otherwise, prominence of an excessively long fibula will result. When the amputation is done improperly so that the muscle retracts too far proximally, the bone end becomes prominent, and excision of the offending bone or reamputation at a higher level is necessary. The muscle should extend only as far as is necessary to provide a padding around the side of the bone. Skin only should cover the end of the stump. When the muscle or the skin is redundant, the fitting is very inadequate, and the tissue becomes edematous and breaks down. Revision of the stump is indicated. "Dog ears" will gradually flatten out under the influence of compression bandaging

Contractures of the stump, particularly in the flexion position, are very disabling, difficult to fit and, in most cases, are avoidable. The muscles become bound down in the position in which they are allowed to rest. A stump allowed to remain in an abnormal position becomes fixed to that position. Energetic early physiotherapy in the preprosthetic period is mandatory. Exercises are designed to strengthen the antagonist muscles, the flexion contracture is stretched, and a neutral position is maintained. If necessary, turnbuckle or wedged casts are used. Surgical treatment to overcome a persistent flexion deformity includes stripping of the muscles and osteotomy followed by special limb fittings. Other causes of flexion contractures include ill-advised longitudinal incisions over flexor surfaces of joints, spastic paralysis and flaccid paralysis.

Involuntary clonic contractions of the stump are frequently seen. This occurs in sudden attacks, mild or severe, and especially in very high-strung individuals. The attack usually occurs during periods of abstinence from the prosthesis, particularly at night, and the individual soon learns to control the jerking motions by firmly grasping the stump. The cause is unknown.

Inadequate circulation within the stump will lead to breakdown of the tissues and infection. Barely sufficient circulation within a stump may not be adequate to cope with friction or pressure of a prosthesis, and soon the tissues break down. This is found most commonly in the below-the-knee stump. Reamputation should be done at a higher level. The diffuse fibrosis found in cases of frostbite obstructs the smallest vessels, and amputation should be done at a sufficient distance from the affected area.

Pain in the stump is caused most commonly by infection or involvement of the nerve trunks. A nerve exposed in an open amputation is productive of much discomfort. The nerve should be cut and allowed to retract and the surface of the stump covered with petrolatum. When a neuroma is encountered, it should be excised. Likewise, a nerve incarcerated in scar tissue should be freed, cut and allowed to retract. This type of pain, as a rule, is not present at rest and is accentuated by pressure or constant movement of the stump Sclerosing solutions and traction on the nerve at surgery should be avoided, careful hemostasis secured, and the nerve end covered with fatty tissue. The incidence of neuroma formation is lessened by these precautions.

The Phantom Limb. Shortly after amputation, most patients perceive a sensation as

putees and a higher mortality rate. The wheelchair existence is in marked contrast with the below-the-knee amputee who is relatively self-sufficient. Early amputation is the only answer to this problem.

Absolute Criteria for Amputation Through the Foot

1. Normal dorsalis pedis pulse
2. Good nutrition of skin
3. Normal skin temperature
4. Normal color on dependency
5. Absence of ischemic pain.

Use of Refrigeration

The limb should be encased in cracked ice up to a level barely above the gangrenous or infected area and kept at a distance from the contemplated site of amputation. The temperature of the effected part is reduced to about 40° C, the protoplasmic activity in that area is lessened, and nerve conductivity is inhibited. Toxic products of tissue destruction are held in abeyance, and their absorption is halted. The result is striking. The fever is lowered, pain is absent, and the patient is converted from a very ill, irrational individual to one who is rational, nontoxic and comfortable. The refrigeration should be continued indefinitely until the condition of the patient is maximum for surgery. Caution should be exercised regarding the placing of the ice. Extreme cold destroys the small-caliber vessel walls. Following its withdrawal, there occurs extravasation of serum, perivascular edema, local hemoconcentration with intravascular obstruction Such tissue responds poorly to surgical trauma, and necrotic wound edges result and necessitate reamputation at higher levels. The use of refrigeration as an anesthetic agent at the operative site is to be condemned. Its value to arrest advance of gangrene and infection, improve the general condition and minimize the shock of surgery is superlative.

PATHOLOGY OF THE AMPUTATION STUMP

Immediate *postoperative bleeding* beneath the wound may develop very slowly as gradual oozing or may be a sudden massive hemorrhage. A ligature may loosen, and the severe hemorrhage may produce sudden pain and swelling. If the amount of blood is minimal, it is treated by aspiration and the application of a pressure bandage. A large amount of blood indicates an openly bleeding vessel, and the operative wound should be opened, the vessel ligated, and the operative incision resutured. Bloody accumulation should be prevented and removed because it leads to scarring and infection. A scarred nonresilient stump lacks resistance to pressure of the prosthesis, and open painful ulceration results.

Superficial *infections* are treated by hot packs, elevation of the stump, chemotherapy and antibiotics. A localized purulent accumulation should be incised and drained followed by plastic repair. Stitch abscesses are most likely to develop about cotton or silk when the wound is sutured under tension. The offending sutures should be removed. When a chronically draining sinus develops, as a rule it is due to a retained foreign body or a nonabsorbable suture. The area should be opened widely down to the foreign body, hot packs applied, and the resulting granulating area excised completely down to normal tissue. Finally, a plastic repair is done. When bone infection is evident, the area should be opened, sequestrectomy performed, and secondary repair accomplished. Anaerobic infection is rare. It is characterized by crepitation, serosanguineous watery discharge, a very characteristic odor and severe generalized toxemia. The area should be opened extensively and an open amputation at a much higher level done

An obstinate *ulceration* may develop about the surgical scar. The causes include inaccurate skin approximation, infection, hemorrhage, a drain which has been left in too long, and circulatory impairment which is due to tight closure, edema of the stump, and excessive compression by the dressing.

Scarred tissues result, and frequent breakdowns occur by friction of the stump in the socket. Treatment is by elevation, rest and hot packs until the infection is controlled and the surrounding skin appears to be normal. Then the scar is completely excised down to the base with exposure of normal-appearing bleeding tissue. Closure of the wound should be effected without tension, if necessary by resection of bone at a higher level.

The *length of the stump* is very important.

stretches forward to reach the toes, thereby rounding the lumbar region as much as possible. Normally, he should be able to reach the toes. Limitation of forward bending is due to (1) hamstring tightness, (2) low back tightness, (3) or gastrocsoleus tightness. Attention directed to these structures corrects the defect.

For General Extensibility. The patient lies supine on a table with legs extended and the pelvis rolled posteriorly so that the lumbar spine is flattened. Next, the arms are extended over the head. Normally, he should be able to touch the table above the head and at the same time maintain a flattened lumbar spine. Limitation (can perform act only by arching the lumbar area forward) is due to (1) tightness of the pectoral musculature, the patient cannot keep the elbow extended and instead must flex it in order to touch the table, (2) tightness of the hip flexors; test by fully flexing the unaffected hip whereupon the involved hip cannot be fully extended to the level of the table (Thomas Test); and (3) hamstring tightness, demonstrated by inability to extend the knee fully while sitting.

Muscle Strengthening Program

Each day the physiotherapist administers heat and massage not only to the stump but also the unaffected extremity. This is followed by exercises which are at first assisted, if necessary, next unassisted and finally resistive exercises. Contractures of joints and the tight muscles are stretched. Unaffected joints are put through full range of motion in order to prevent contracture. This program should be instituted immediately after surgery and continued throughout the preprosthetic and the prosthetic periods. The circulation of the stump should be aided by various means, including Buerger's exercises, preganglionic sympatheticolytic drugs, such as Priscoline, and an ounce of whiskey several times a day.

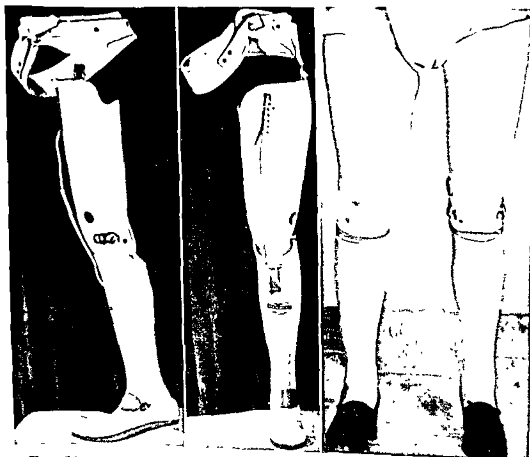


Fig 546. (*Left*) Side view and (*center*) front view of conventional prosthesis for an above the knee amputation. (*Right*) The suction socket limb. (The American Limb Co.)

though the lost portion of the limb were still present, but this feeling gradually disappears. However, the term "phantom limb" is applied to that condition in which the sensation is persistent, painful and annoying. Relief may require the use of morphine. A source of irritation in the stump, such as a neuroma, is the most plausible explanation for the phenomenon However, the many types of treatment, including resection of the neuroma, sympathectomy, procaine block, posterior ramisectomy, chordotomy, etc., constitute mute evidence of frequent failure.

Hyperesthesia of the stump is another annoying symptom which is difficult to control Reamputation results only in reproducing the symptom at a higher level.

Elastic bandaging is done postoperatively to reduce the edema and to shrink and mold the stump prior to fitting for a prosthesis. Improper bandaging will result in infolding of the skin with deep creases at a right angle to the scar, pressure sores, markedly swollen edematous tissues distal to a tight constricting turn of bandage, and poorly shaped stumps which are difficult to fit. The bandage should be applied and reapplied with even pressure throughout, care being exercised to avoid excessive pressure and constricting turns of the bandage

Blisters, swelling and pain due to pressure sores produced by a poorly fitted prosthesis are avoidable The proper application of an artificial limb requires the constant conscientious observation and co-operation of the surgeon and limb-maker. The inner surface of the socket should be smooth and devoid of abnormal elevations. Any impediment to the venous flow within the socket or by the corset edge where it compresses the popliteal space should be eliminated The posterior rim of the socket may press on the popliteal vessels. The distal end of the socket in a below-the-knee limb should be loose. In the above-the-limb prosthesis, the anterior rim of the socket may compress vessels in the femoral triangle. The suction socket limb is held on the stump by the negative pressure within the socket. This pressure when excessive produces swelling and breakdown of the tissues, particularly in a stump with poor circulation. Gradual adaptation of the stump to the limb may obviate this difficulty. The soft tissues gradually toughen and resist the negative pressure. Pressure sores, especially over bony prominences, may be avoided by proper excavations on the inner wall of the socket and by use of thick wool stump socks.

PHYSICAL REHABILITATION OF THE AMPUTEE

Before the amputee can resume normal living activities and become self-sufficient, he must be reconditioned physically and taught to use the artificial limb. Strengthening of muscles, attainment of balance, and repeated practice and development of habit patterns are necessary prerequisites to full rehabilitation. This demands constant surveillance by orthopaedic surgeon, physiotherapist and limb-maker.

Determination of Deviations from Normal Posture

Normal Posture. This is determined by the plumbline tests. A plumbline is dropped from the ceiling and approximated to the mid-line of the body in the anteroposterior and lateral positions.

Anteroposterior Test. The plumbline passes through a point just anterior to the lateral malleolus, the knee joint posterior to the patella, the hip joint, the midabdominal region laterally, the shoulder joint and the lobe of the ear.

Lateral Test. The plumbline passes through a point midway between the medial malleoli, the coccyx, the sacrum and the spinous processes. Good posture is essential to balanced walking. Any deviations are clues to the injurious factors which are operative For example, contracture of the hip flexors on the stump side may cause forward knees, forward hips, marked lordosis with rotation of the pelvis, forward displacement of the shoulders and the head A front view shows the body to be displaced to one side because a disproportionate share of the body weight is borne on the normal leg Correction of flexion contracture, posture exercises and careful instruction correct the defect, provide better balance and reduce fatigue when walking.

Examinations

For General Flexibility. The patient sits on a table with the legs fully extended and

for several inches at the top only; elsewhere there should be contact only between socket and anterior surface of stump. The shape of the opening at the top is unimportant.

Main problems in fitting and use:

1. Whip of knee in swing phase with rotation of the leg at the heel strike. The cause is malalignment of the knee axis.

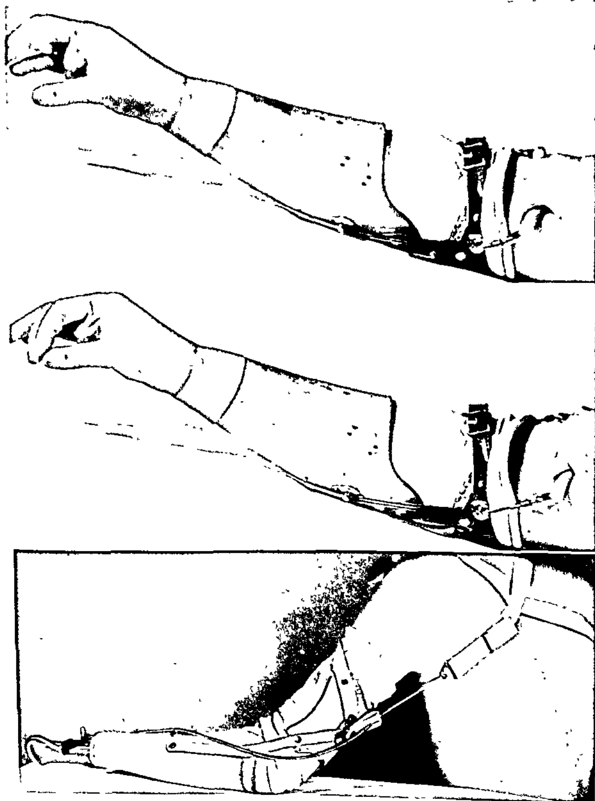


FIG. 547. (*Top*) Prosthesis for the cineplastic stump. Motor power is supplied by the biceps muscle. Relaxed position. (*Center*) Contracted position. (*Bottom*) Prosthesis for below-elbow amputation stump. The power for the hand mechanism is supplied by the opposite shoulder through a cable. (The American Limb Co.)

Bandaging

The patient is instructed on the proper procedure of bandaging the stump with an elastic bandage to promote uniform shrinkage. Various physical agents may be used to aid the final stump, preparing it for the prosthesis These include whirlpool baths, ultraviolet rays and procaine ion transfer. In this last procedure a solution of procaine hydrochloride in 95 per cent alcohol is applied over the end of the stump, and from 3 to 5 milliamperes of constant current is applied for about 15 minutes. This procedure effectively relieves pain.

Fitting of the Prosthesis

The stump must be well healed, the skin of good thickness and texture, circulation adequate, scars away from points of pressure, and the muscles strengthened by energetic resistive exercises. The length of the stump should be adequate for the desired limb. The limb should be fitted by an experienced artificial limb manufacturer and under the supervision of the attending surgeon. At first the limb should be worn for short periods of time, and each successive period is gradually increased in order that the patient may become accustomed to the feel of the new object.

Balancing Exercises

Balancing exercises are performed in front of a mirror so as to promote co-ordination of all muscles in both extremities. The patient is encouraged to stand erect in a proper posture, which is a prerequisite for good balance. The patient slowly squats down, holds the position for a few seconds, then assumes the erect position. This is repeated a number of times, and the depth of knee bend is increased. Particular attention is paid to the gluteal muscles, the tone of which is very necessary for maintaining the erect position. While standing on the affected extremity, the opposite extremity is moved forward slowly, sidewise and backward, and the procedure is repeated with increasing rapidity. The uninvolved hip may be strengthened likewise. The patient stands between parallel bars with the feet placed about 2 feet apart and shifts his weight from one leg to the other to get the feel. The weight is borne on the prosthesis, and the individual alternately steps forward and backward with the normal leg. After the sensation of stability is attained, the patient should next *practice walking* in a straight line with the heels close together. A bilateral amputee should always walk with the legs apart for better stability. The normal walking rate is approximately 60 steps per minute. Finally, a variety of walking movements performed in everyday activity is practiced. This includes climbing stairs, descending stairs, turning about, sitting down, rising from a chair, bending for objects on the floor, and climbing and descending from a curb.

Some limb manufacturers provide the facilities and the supervising individuals so that the amputee may perform these various necessary motions and the use of the limb be better studied and adjusted for a proper fit.

Suction Socket Prosthesis[1, 2]

This type of prosthesis is one held on by suction and close anatomic fit and is most suitable for above-the-knee amputations. In contrast with the older conventional prosthesis, it eliminates the hip joint and pelvic belt or shoulder harness, it permits freer rotatory motion about the hip, it eliminates piston action of the stump in the socket, permitting greater toe clearance and smoother gait; no stump sock is needed; and the adductor roll is reduced or eliminated. In essence, the socket is closely fitted to create negative pressure during the swing phase and positive pressure which expels air through a flap valve during the stance phase. The tight fit is applied at the upper 2½ or 3 inches and along the anterior wall of the socket, the remainder of the stump hanging free. Some weight-bearing is sustained on a sloping ischial seat at the posteromedial border of the opening. A groove in the anteromedial wall accommodates the adductor tendons. The outer edge of the opening is at a slightly higher level than the medial edge.

The two main points in construction are: (1) the ischial seat should be wide, flat and well toward the medial side directly beneath the tuberosity; and (2) the socket should be barrel-shaped, snug-fitting circumferentially

[1] Mazet, R, Jr., McMaster, P. E, and Hutter, C. G.: Analysis of 124 suction socket wearers followed from 6 to 55 months, J. Bone & Joint Surg 33A 618, 1951.

[2] Von Werssowetz, O F.: Above-the-knee suction-socket prosthesis, J Bone & Joint Surg 24A:731, 1952.

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2. Rotation of leg at heel strike or toe off. Causes are (1) too tight a fit in the adductor region; (2) incorrect knee alignment; (3) ankle joint in wrong axis.

3. Medial lateral instability during stance phase. Causes are: (1) incorrect alignment of foot; (2) malaligned knee.

4. *Swelling and discoloration at end of stump* The cause is constriction above the level of edema. With use, the muscles of the stump hypertrophy and contribute to the choking. Treatment consists of elevation, massage and compression bandaging.

5. Skin irritation in adductor region. Relieve pressure in the adductor channel.

6. Perspiration of stump. Apply 20 per cent aluminum chloride to the stump every 3 or 4 days.

7. Fungus in socket. Wash socket interior with 2 per cent formalin followed by alcohol at weekly intervals.

The valve is a springless flap valve which expels the air on standing and closes during the swing phase when a negative pressure of ½ to 1 pound per square inch develops. It remains open in the sitting or reclining position, permitting a partial air exchange in the socket.

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